PEDIATRIC NEUROSURGERY

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DEDICATED TO

SIR GEOFFREY JEFFERSON

and

WILLIAM VERNON CONE

Two Neurosurgeons
who have inspired many

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Preface

THIS VOLUME, delineating pediatric neurosurgery from adult neurosurgery in the miniature, is prepared for those physicians who care for these neurologically ill patients.

The study of and interest in neurological disease in pediatric patients has become more intense in recent years as evidenced by the subspecialization of some neurologists and neurosurgeons in this subject. It was felt that there was needed a survey of neurosurgical diseases in pediatric patients, in which opinions represented a cross-section of the country. This volume has been prepared by twenty-four contributors from all parts of the United States. Contributors include neurosurgeons practicing in general hospitals, large private clinics, university hospitals, and such medical specialists as are closely associated with these patients as the pediatrician, anesthetist, plastic surgeon, neurologist and electroencephalographer. It is our belief that the infant or child afflicted with a surgical disorder of his nervous system requires the combined efforts of the pediatrician, neurosurgeon, and other ancillary medical therapists. Because of the advances in diagnostic and therapeutic techniques, fluid balance, anesthesiology, patient care, and rehabilitation, each specialist has his own contribution to make to the general welfare of the patient. The majority of youngsters in this country are cared for by the general practitioner and, therefore, he too shares in the advancement of pediatric neurosurgery for he is most frequently the first to see the patient with a neurosurgical lesion. All of this reflects favorably on the dissemination of new information from specialist to specialist, and from specialist to general practitioner.

This volume is divided into two parts. The first part, comprising seven chapters, deals with general considerations related to pediatric neurosurgery, mostly prepared by the ancillary specialists except for two chapters by the editors. The second part, comprising 14 chapters, deals with specific neurosurgical entities, for the most part under classical etiologies and written by neurological surgeons.

We are indebted to Doctor Hendrik Svien of the Mayo Clinic who spent a considerable amount of time and energy in helping plan and outline this volume at its conception. Our sincerest appreciation to Mrs. Ira Jackson for the many hours spent reading proof from the earliest drafts

through the final page proof. The original drawings were executed by: George Newman of Galveston, Texas—Chapters II and IX; Yun Yong of Philadelphia—Chapter XV; K. Mackay of Atlanta, Ga.—Chapters II and XVI; M. Guthrie of Memphis—Chapter X; T. Marshall of Minneapolis—Chapter XII; E. Hoff of Detroit—Chapter XVIII; and Dr. G. Eade of Galveston—Chapter VI. The photography was done by those associated with the various authors in their universities or hospitals.

We are grateful to our contributors who sacrificed much time, energy, and expense in preparing their chapters—and for their patience and understanding in carrying out revisions. Any shortcomings in this volume are the sole responsibilities of the Editors.

To Charles C Thomas, Publisher—we are most appreciative for their encouragement, advice, and cooperation throughout the preparation of this volume.

I.J.J.
R.K.T.

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PEDIATRIC NEUROSURGERY

PART I

CHAPTER I

Introduction

Raymond K. Thompson and Ira J. Jackson

PEDIATRIC NEUROSURGERY is that surgery of the entire nervous system between the time of birth and the age of fourteen. At this time, the cranial sutures become fused and the treatment of the problems and diseases of the nervous system of childhood ceases and shades over into the treatment of the diseases of the adult nervous system. Pediatric neurosurgery is more than adult neurosurgery in the miniature. The small size coupled with the difference in anatomy and difference in physiology of infancy and childhood warrant that this phase of neurosurgery be considered separately from that of the adult.

Anatomical Considerations

The average circumference of the skull at birth is approximately 35 cm. During the first year an increase of approximately 10 cm. in circumference occurs. This growth occurs at the rate of about 1.25 cm. a month for the first six months, and after that approximately .75 cm. a month up to the first year. During the second year of growth the skull increases about 2.5 cm. From the period of two years to five years the skull increases about 4 cm. From this point up to the fusion of the sutures the skull grows at the rate of about .5 inch in circumference during each five-year period. The rapidly growing skull is made flexible by the presence of the unfused suture lines and the fontanels between various bones. This flexibility allows for distention of the cranial cavity with small increments of increased intracranial pressure, whereas the adult skull does not permit such enlargement. Consequently, as increasing pressure develops during this period of growth, the suture lines are separated and the fontanels bulge. This lessens the force of increased intracranial pressure. It is abnormal for the cranial sutures to be separated after birth. It is not until the sixth month that the cranial sutures become immobile and, at times, mobility can be demonstrated until the ninth month. Following this, the suture lines are held in apposition by connective tissue until their eventual fusion at the age of approximately fourteen years.

The fontanels are present at birth. The posterior fontanel between the parietal and occipital squamae usually becomes obliterated by the eighth week. The anterior fontanel usually becomes occluded between the fifteenth and the eighteenth month. At one year, the tip of the finger can be admitted into the anterior fontanel, and its measurement usually is 2 cm. An anterior fontanel which is open at the end of the second year of life is considered definitely abnormal. During the early period of development, when the fontanel is open, pulsation of the intracranial contents can be seen under the normal circumstances, while in the presence of increased intracranial pressure, the fontanel loses its pulsation. Under normal circumstances, the fontanel is slightly depressed when the child's head is held in the sitting position, whereas it fills out when the child lies down. When the child is held in the erect position, and the fontanel no longer sinks beneath the surface, increased intracranial pressure is present.

Encased in the skull and meninges is the developing brain which is undergoing marked developmental changes. The most carefully studied developmental changes are those of myelination of the nervous system. It is simultaneous with myelination that function begins in various parts of the nervous system. Both the brain and spinal cord have become progressively more myelinated until at approximately two years of life this process becomes complete. Concurrent with this myelination occurs also a growth in neuron and glial cells, as well as the vascular components which supply the nervous system. It is, therefore, of little wonder that at birth the brain is quite gelatinous in consistency and must be handled with infinitely more care than even the most meticulous manipulations of the adult brain.

The spinal cord, under normal circumstances at birth, extends to its caudal termination at the lower border of the 2nd lumbar vertebra. Because of the more rapid growth of the osseous components of the spinal column, this level gradually rises until, as an adult, the caudal end of the spinal cord ends just above the border of the body of the 2nd lumbar vertebra.

Physiologic Considerations

During infancy and childhood, there are many physiologic differences between the child and the adult. However, consideration of circulating blood volume is the prime physiologic variant of interest to the neurosurgeon. It is with blood loss that the pediatric neurosurgeon must primarily concern himself. In the adult, the blood reservoir of the body contains about 5.5 liters of blood, or, more accurately, about 78 cc. of blood per kilogram of body weight. This blood volume is much smaller in infants and children and does not reach the normal adult figure until

approximately sixteen years of age. It is easy to visualize, therefore, the extremely small blood volume, beginning at birth and through the early years of life. The approximate totals of circulating blood volume are: at birth, 268 cc.; at three months, 577 cc.; at one year, 741 cc.; at two years, 875 cc.; at five years, 1,216 cc.; at ten years, 2,184 cc.; at fourteen years, 3,120 cc., or just slightly more than half the blood volume of the adult. Since blood loss in intracranial and spinal procedures can be rather extensive, it is quite obvious that even a small amount of blood loss materially reduces the blood volume in the child, and disastrously reduces the blood volume in an infant.

Body temperature must also be carefully watched in pediatric patients. Since the body surface of infants and children is relatively large, a disastrous amount of heat can be lost from the child under general anesthesia. On the other hand, excessive heat from blankets and overheated operating facilities can generate tremendous hyperthermia, since only a small caloric exchange is necessary to alter the body temperature. For this reason, the body temperature should be measured on pediatric patients repeatedly during any prolonged operative procedure and appropriate measures taken to maintain correct body temperature.

Total oxygen intake of an infant or child is subject to wild fluctuations of body oxygen content with only minimal changes in altering factors. An obstructed airway or a marked anemia can produce a hypoxic state in a child much more easily than if the total body oxygen were of larger mass. For this reason, meticulous care to the maintenance of a free airway during anesthesia and attention to blood loss and preoperative anemia are of major concern.

Technical Considerations

Since operating rooms are devised for adult surgery, modification of the adult facilities creates a number of technical problems, and actually the diminutive size of the patient in pediatric neurosurgery involves altering many of the existing facilities to suit the child. In preparing an infant for surgery, positioning and immobilization of the body on an adult headrest and operating table can be accomplished by the use of adhesive tape and blankets, or towels, which have been rolled into appropriate shapes. The ordinary operating room headrest can be filled in with Dakin pads until the opening in the headrest approaches the size of the infant being operated upon. These pads can then be held in position by a roller gauze bandage applied around them. It is possible to roll a towel and then shape it into the form of a doughnut, which, in turn, can be bandaged so as to produce an appropriate-sized, doughnut-shaped, rest for the

infant's head. Under general anesthesia, immobilization is not as great a problem as when working under local anesthetic drugs. Under these circumstances, the child can frequently be held properly in position by bandaging the whole infant to a circumcision board. A Dakin pad, cut to size and wrapped around the wrists, can be easily fixed to the side of the table with gauze bandage. Adhesive strapping across the hips, or across the scalp, will at times materially help with immobilizing the operative part. At times it is possible that fixing the infant's diaper and jacket to the drapes of the operating table with safety pins will aid in immobilization of the child. A word of caution might be said regarding heavy drapes resting on pediatric patients. The use of an overhead table allows the drapes to be attached to them, so as not to rest upon the child, and this gives the anesthesiologist freer access to the child.

In the preoperative period, a cutdown should be performed so that the administration of blood and fluids can be carried out throughout the operative procedure as needed. The saphenous vein lying just above and ventral to the medial malleolus is the vein which is usually used for this purpose. With the child restrained, the medial aspect of the ankle is made sterile, after the leg has been fastened to a sandbag by means of adhesive strips. This prevents the child from excessive movement of the extremity which will at times dislodge the cannula. The area is draped off and a small incision is made transverse to the course of the vein, just above the medial malleolus, after the infiltration of a small amount of procaine. The vein is easily identified in almost all children, even those which are marasmic or badly dehydrated. After the vein has been identified, a silk tie is placed under it and a small nick is made in the vein with an iris scissors. Suitable-sized polyethylene tubing can then be threaded into the vein for a distance of several centimeters, and an appropriate needle attached to the free end of the polyethylene tube. Saline can be allowed to drip slowly through this, to maintain its patency. A silk stitch is placed on either side of the polyethylene tube, closing the incision. A dressing is applied and strapped in place with adhesive tape and the polyethylene tube is circled around and strapped with several bands of adhesive tape, to insure that the cannula is not dislodged by a sudden jerk on the tubing. This may be left in place for several days and removed simply by pulling the cannula from the vein and exerting pressure on the wound for several moments.

The control of the child's temperature on the operating table can be simply measured by a rectal thermometer read at regular intervals. There are available, however, thermocouple types of constant recording equipment, so that body temperature may be graphed or read repeatedly from

a meter. The constant observation of temperature during lengthy neurosurgical procedures cannot be stressed too much, as it will immediately point out the rapid accumulation of heat under excessive dressings or the rapid loss of body heat due to irradiation from the skin in a cold operating room. An electric blanket is ideal, under the child, if the temperature of this blanket can be accurately maintained. However, the use of hot water bottles is simple and very efficient.

The vascular system of the child should be carefully watched, and it has been found very helpful to have the bell of the stethoscope strapped to the chest wall so that the cardiac beat can be constantly heard by the anesthesiologist. A small infant blood pressure cuff is also mandatory during neurosurgical procedures of any magnitude.

Instruments for pediatric neurosurgery are being devised daily, in that the adult-sized instruments are at times clumsy and dangerous in handling the excessively delicate tissues. The use of the small size towel clip materially reduces the weight of the drapes on the child. A simple suture, fixing the drape to the child's skin, is preferable to heavy towel clips. The use of the small knife-blade (No. 15 Bard-Parker) allows the surgeon ease in incising diminutive structures. The scissors of eye surgery are ideal for the handling of children's tissues. Both the blunt and pointed scissors are available and are of a size to allow easy manipulation in cutting tissues. Several instrument companies have available now a long, fine needle-holder, with specially prepared jaws, which allows the easy manipulation of small French needles, which are actually ideal for suturing infants' tissues. The thin skull bone of the infant can be frequently cut with the adult operating scissors, allowing rapid elevation of an osteoplastic flap. The use of curved or straight mosquito clamps replace the large straight clamps which are used on the adult, for everting the galea for hemostasis of the scalp. The rubber-coated Frazier-Bucy suction tip allows the surgeon's assistant to have coagulating current as well as suction available and easily manipulated with one hand. The value of extra length cautery wires and extra length suction tubing, as well as extra length cable from the cautery to the foot-pedal, cannot be emphasized enough. This allows the cautery and suction apparatus to be placed far away from the operative and anesthetic sites and permits easy mobility around the operating table by the non-sterile attendants. Diminutive brain spatulas can be easily fashioned from sheet brass, to the size and shape required by the problem presented. With the use of these and other small instruments, the ease and speed of the operation can be greatly facilitated.

It is felt that consideration of the specific operative technique involved in care of the child is best handled in this type of text book by allowing

each author to detail as necessary the procedures done under each of the chapters. For example the technique for craniotomy is dealt with under the chapter of brain tumors while that of laminectomy is under the chapter of spinal cord neoplasms. The details of shunt procedures are described under the heading of hydrocephalus and the technique specific to the treatment of the subdural membrane is under the appropriate chapter. The reader therefore will refer to the individual chapters for the details of the surgical procedures in the specific cases.

CHAPTER II

Diagnostic Techniques

Ira J. Jackson and Raymond K. Thompson

THE VARIOUS technical procedures utilized in the diagnosis and at times in the treatment of various neurological and neurosurgical disorders as presented in this book are described only as they pertain to the infant and child differing from the adult. Only the technique itself is discussed leaving the interpretation to follow in the various chapters which are concerned with these methods under specific conditions.

Lumbar Puncture

Lumbar puncture in the infant can be easily performed in the majority of cases if the infant or child is held in a relatively fixed position. The puncture can be carried out with the patient either in the lateral position or sitting up. Utilizing the lateral-horizontal position in the infant or restless child, a rolled sheet is placed around the neck at one end and behind the knees at the other end, pulling the two ends together forming a figure eight thereby causing acute flexion of the spine. With the sheet so placed and firmly held by a nurse the patient can be maintained in a fairly fixed position. The patient's back should be brought to the edge of the table and kept as perpendicular as possible to the table. It is always much easier doing such a puncture on a table rather than a bed or crib.

Another popular method for holding an infant in a fixed position for a lumbar puncture can be easily accomplished by the assistant placing the index fingers of either hand around the lower part of the humerus and then hooking the fourth and fifth fingers under the knees of the infant with the infant sitting facing him, allowing the head to fall forward on his arms. Holding an infant in this manner in the sitting position, the assistant can lift the child up, turn the child on the side, or maintain it in a relatively rigid position so that the physician doing the lumbar puncture can perform this with minimal mobility of the lumbar spine.

Gently but forcibly indenting the skin over the interspinous ligament in the lower lumbar region produces a localized ischemia and partial anesthesia. If firm pressure of the fingernail is maintained on the skin

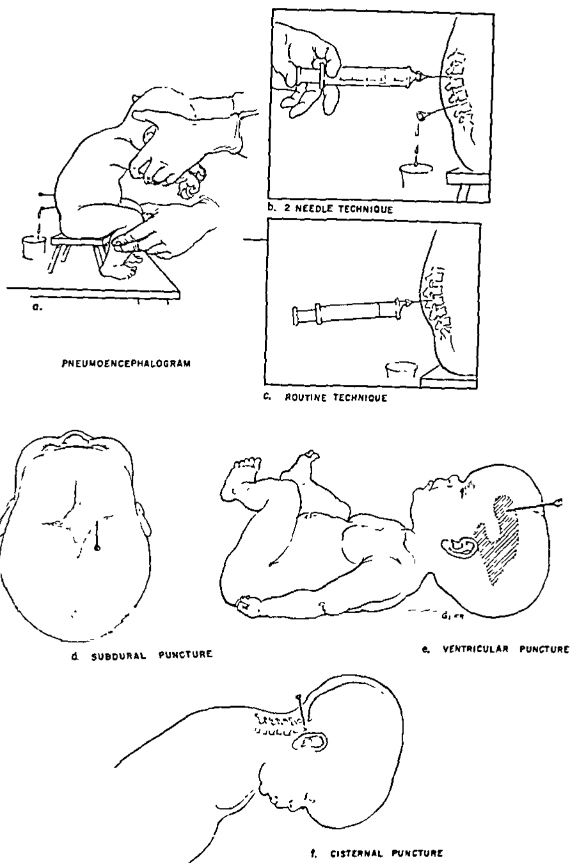


FIGURE 1. (a) Pneumoencephalogram performed with the infant or very young child held on small stool utilizing the two needle method. Without a stool or large sand

for a period of thirty seconds, the puncturing needle can be rapidly introduced through the skin with minimal pain to the infant.

Local anesthesia is not necessary in infants but should be used in children just as for the adult. The infant can be given a bottle of glucose and water or a sugar nipple during the procedure so that introduction of the needle can be performed without stimulating him too much.

Pediatric lumbar puncture needles are available in most hospitals and should be used. These needles are identical to those for adults except are considerably shorter in length. If no such needles are available, the regular adult needle may be cut off so that it measures only 3 to 3½ inches long. Either an 18 or 20 gauge needle is preferable. A number 22 gauge needle has too narrow a lumen requiring too long a time for the fluid to flow out and be collected or to measure the pressure. It is wise for the person performing the lumbar puncture to reflect on the very short distance required to penetrate the subarachnoid space in small infants. If one mentally pictures the size of the lumbar vertebrae, the all too frequent, too deep, introduction of the needle will be prevented. After introducing the needle through the skin and into the interspinous ligament, it is well to release the needle and note the direction it is taking. This simple maneuver frequently allows for proper direction of the needle. With the child in the sitting position, the correct direction of the needle is more easily seen. Once the spinal needle is in place in the subarachnoid space, the assistant's grasp need not be changed to lay the child down on its side, maintaining immobilization. The quantity of spinal fluid removed in the infant and child is the same as in the adult. Following the lumbar

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bag, the hubs of the spinal needles will be too close to the table top since the lower lumbar spine in the infant is but inches away from the buttocks. With the two needle method, both needles must contain the same sized lumens, and excellent flow of fluid present at both needles. Air may then be introduced at a rather rapid rate with a large syringe through the upper needle while the fluid will be spurting out of the bottom one, and the entire exchange of air and spinal fluid taking but a few minutes. (b) Close up of (a). (c) Pneumoencephalogram performed by single needle and small amounts of air introduced through an eccentric syringe in exchange for an equal amount of spinal fluid each time The exchange may be done in 2-5 cc. increments. The eccentric syringe allows one to introduce air which stays on the top of the barrel in the syringe while the spinal fluid is collected in the bottom of the barrel. (d) Subdural tap. Needle introduced at lateral angle of anterior fontanelle in tangential fashion (e) Ventricular puncture. Needle introduced through lateral angle of anterior fontanelle in line with the pupil and perpendicular to the skin or pointing slightly posterior. (f) Cisternal puncture. The point of the needle is placed on the rim of the foramen magnum and gently and slowly slipped inferiorly to pierce the dura and enter the subarachnoid space.

puncture, the infant or child may resume his previous activities without restrictions because of the tap.

Cisternal Puncture

This is most easily accomplished with two assistants; one holding the child in the sitting position, and the second holding the head. The occipital region and upper cervical region is cleaned and the level of the foramen magnum identified by palpation. The point of the needle is first directed against the rim of the foramen magnum and then gently slipped around the edge of the bone and into the cisterna magna. The two assistants can then allow the child to be placed on its side without changing the relative positions of the head and neck. No anesthesia is necessary in the infant while local anesthesia is used in the children. Under unusual and rare circumstances, general anesthesia can be utilized.

Subdural Puncture

A subdural puncture can be done through the skin only as long as the anterior fontanelle remains open, which automatically makes this a procedure for only the very young patient.

The infant is "mummified" by wrapping his body and extremities securely in a sheet (Fig. 1). This will permit the assistant to hold the head firmly and eliminate annoying struggling. Scrupulous aseptic technique is imperative. The coronal area of the scalp should be properly shaved and prepared with ether and soap and water. It is not necessary to use local anesthesia because it will frequently cause as much pain as actually introducing exploratory needle. The needle most commonly used is a 1 inch 18 or 19 gauge, with a sharp short beveled tip containing a stylet. Short needles are preferable, because they are more easily managed and one is not as likely to penetrate the cortex. The puncture is carried out at the lateral angle of the fontanelle to avoid the superior longitudinal sinus. As the needle penetrates the fontanelle membrane, formed by fusion of the dura and periostium at the bony margins, one usually feels a slight "pop" as the subdural space is entered. The needle should be introduced at a mild angle to the scalp to avoid continuity of drainage channels and subsequent "weeping" after the puncture has been effected.

When carried out initially, bilateral punctures should be performed. It should be stressed that under normal conditions no fluid is obtained from the subdural space. If only a few drops to a cubic centimeter of clear colorless fluid is obtained, the needle has probably penetrated too deeply and is in the cerebral subarachnoid space.

Aspiration of the needle in the subdural space should not be done and is unnecessary because of the possibility of sucking up cerebral tissue. If the needle is in the subdural space in a patient with a subdural hematoma

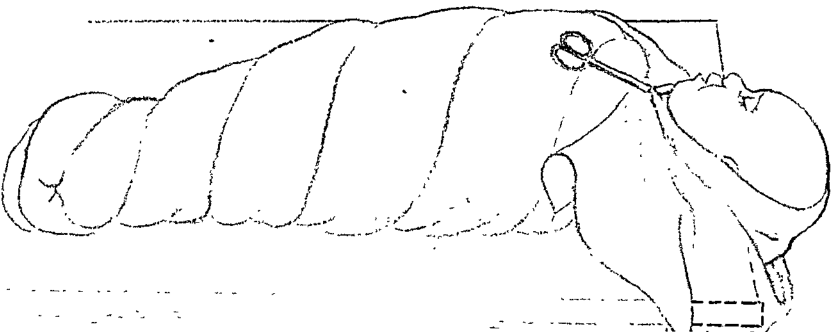


FIGURE 2. Method of holding infant (mummifying) in order to perform a subdural or ventricular puncture. (*Courtesy*, Dr. Robert Mabon.)

or effusion, the fluid will flow freely and quickly with an 18 or 19 gauge needle. Objection to a 22 gauge needle is that the flow of fluid is so slow one can hardly be certain the needle is in the proper space.

After the needle has been withdrawn, a very small piece of gauze saturated with colloidion is used to cover the punctured hole in the skin rather than a piece of adhesive tape over a piece of gauze. This small dressing permits unobstructed observation of the size and pressure of the anterior fontanelle and also is easier and less traumatic to remove.

Ventricular Puncture

In order to puncture the ventricle of an infant with an open anterior fontanelle, one proceeds initially in a manner similar to that described under Subdural Puncture.

A 20 gauge pediatric lumbar puncture needle is used and cautiously directed into the cerebral hemisphere at right angles to the scalp in line with the pupil of the ipsilateral eye. The depth at which the ventricle will be entered will obviously vary with the size of the anterior horn of the lateral ventricle. The stylet is first withdrawn after the needle has punctured the dura to see if there is any subdural fluid. The stylet is replaced, and the needle is introduced about 1 centimeter before checking to see if the ventricle has been punctured. The needle is introduced a ½ cm. at a time until fluid is obtained. However if the needle is in the brain about 3 cm. without entering the ventricle, the ventricles are probably normal or only slightly enlarged in size and should be withdrawn and reentered altering the direction so that the point of the needle is more posterior and medial. Some surgeons believe that if the ventricle is not entered into after the third attempt, it should be temporarily abandoned.

The amount of fluid withdrawn will depend upon the reason for doing the tap.

Following the completion of the puncture, a dressing is applied as described for subdural punctures. The infant may then resume the normal activities with no restrictions.

If the anterior fontanelle is small and barely open, it is not advisable to perform a ventricular puncture through this route for fear of striking the sagittal sinus or a large adjacent vein. If the intracranial pressure is increased and the anterior fontanelle closed, the coronal suture may be sufficiently open to admit a needle for tapping the ventricle.

In the child, a ventricular tap is carried out as in an adult requiring a burr hole.

Pneumoencephalography

Air or oxygen is introduced into the ventricles, cisterna, and cerebral subarachnoid spaces through the lumbar subarachnoid space by means

of a lumbar puncture. The patient is held in the sitting position while the air is injected. The infant can be held as described in performing a lumbar puncture in the sitting position, while the child is positioned upright as an adult.

The procedure may be done under local or general anesthesia regardless of age, and is dependent only upon the personal preference of the neurosurgeon.

Once the needle is properly inserted, the initial pressure is measured. It is the height of the fluid column in the manometer in relation to the vertex that is important as to whether the pressure is or isn't increased—and not the actual millimeters of pressure. The intracranial pressure is considered to be elevated above normal if the fluid level in the manometer is above the top of the patient's head. However, if the patient has been straining, or if the intubation was difficult, or if the head is laterally flexed compressing the jugular vein, the pressure will be increased. Most neurosurgeons perform the pneumoencephalogram in the absence of clinical increased intracranial pressure.

Five ccs. of air are slowly introduced before any fluid is removed. Then equal amounts of fluid are replaced with air—usually in 5 cc. increments. Air is injected until no more fluid comes out. However, a small pneumoencephalogram may be performed in which only a total of 20-30 cc. of air is replaced.

The exchange of air and spinal fluid may be carried out by the use of two spinal needles—one placed at a level below the other. When this technique is used, the air is rapidly introduced in large increments through the upper needle while the fluid is ejected through the bottom needle at a rate equal to the introduction. Air is injected until it comes out of the lower needle. The entire exchange takes only a few minutes this way.

An 18 to 20 gauge needle is utilized for pneumoencephalograms. When using the two needle technique, both needles must be the same gauge and preferably No. 18.

The taking of the x-rays is a radiological problem and will not be discussed. A variety of positions are used in order to completely visualize the entire ventricular system.

The post pneumographic symptoms as headache, nausea, and vomiting are treated symptomatically as in the adult.

Ventriculography

A ventriculogram is done when the intracranial pressure is thought to be elevated. In the case of the infant, it is performed as described for a ventricular puncture through the lateral angle of the anterior fontanelle. Fluid is exchanged for air in 5 cc. increments with the infant held on its

side so that the head is in the lateral position. The air is introduced on the side closest to the table, and since air rises it will fill the opposite ventricle first. The procedure may also be carried out with a needle in each lateral ventricle—the air being injected in the bottom one until it comes out of the top one. If the ventricles are considerably dilated a complete exchange should not be done because it is rather shocking to the patient. Usually 100 cc. of air will produce satisfactory information in the large ventricular system when careful positioning is done filling each part of the ventricular system with air. The "bubble technique" consists of putting in 50 to 100 cc. of air in ventricles that hold perhaps 2-4,000 cc. of fluid.

In some clinics, large amounts of air are replaced—even as much as a 1000 cc. After roentgenograms have been taken, the air in the ventricles is replaced with an artificially prepared cerebrospinal fluid. A needle is placed in each lateral ventricle and with the head in the lateral position the fluid is allowed to drip into the bottom needle at its own rate causing all the air to escape from the needle in uppermost ventricles. The artificial spinal fluid is given until fluid finally comes out of the top needle.

The ventriculogram is done without anesthesia in the infant though he is given a sugar nipple or a bottle of glucose to suck. The child is done either under local or general anesthesia.

The post-operative complaints are treated symptomatically as in the adult. However, there are occasions when the ventricle will have to be tapped to release air when symptoms of sudden increased intracranial pressure occur.

In the infant with a closed fontanelle or in the child, the ventriculogram is performed as in the adult.

Myelography

The visualization of the spinal subarachnoid space in infancy and childhood is similar to that of the adult except that a smaller quantity of radiopaque oil is used. The lumbar needle is introduced as described under the technique of Lumbar Puncture. A No. 18 gauge needle is always used because of the heavy viscosity of the oil. One half to one cc. of radiopaque oil is frequently all that is necessary for careful visualization of the spinal subarachnoid space in the infant, while in the child, 3 cc. is often used. If evidence of spinal block is present, the oil should not be removed through the needle. When the examination is within normal limits, the oil can be centered under the lumbar puncture needle and, with extremely gentle traction on the plunger of a 2 cc. syringe, the oil can frequently be completely removed. In order to maintain the lumbar puncture needle in place during myelography, the myelogram should be performed in the

x-ray department, on the tilttable. Once the oil has been introduced into the subarachnoid space, the child can be gently rolled on its stomach and strapped on the x-ray table with strips of adhesive tape, to maintain it immobile during the x-ray maneuvers. At times, introduction of the oil through the cisternal route is advisable where free communication with the subarachnoid cannot be established with a lumbar puncture needle or where the upper border of a block of the subarachnoid space needs to be visualized.

When a cervical tumor is suspected, it is advisable to plan to carry out the laminectomy immediately afterwards because on occasion acute progression of symptoms has taken place immediately following the myelogram.

The technique is done under local anesthesia except when the infant or child is unmanageable and then general anesthesia in the form of an intravenous preparation is administered.

Arteriography

Arteriography in pediatric patients practically always must be done under general anesthesia with the strongly recommended use of an intraracheal tube. Hypaque®* in 35% concentration is now most frequently used throughout this country. Previously, other contrast medias were used as Urokon, diodrast, and lipiodol. A sensitivity test by the intracutaneous or intravenous method is always performed before performing the arteriogram. The percutaneous method is most frequently employed especially by those surgeons with considerable experience in pediatric carotid arteriography. If the common carotid artery cannot be punctured through the skin, it is surgically exposed in the neck. When the percutaneous method is done, the skin of the neck is prepared with soap and water and then the area is painted with mecresin or some such material. If the artery is directly exposed, a regular surgical preparation is employed. The type of needle utilized in puncturing the artery varies considerably with the many surgeons. Some believe the 18 or 20 Cournand needle is most satisfactory. Others use a short 18 gauge spinal needle. Some prefer to use a plastic needle in the infant in which there is actually a double needle—the inner metallic one acting also as a stylet for the outer plastic one. There was recently described a method whereby the temporal artery was surgically exposed and a small plastic catheter passed down the artery a specified distance entering the common carotid artery.

The amount of contrast material injected for each projection also varies but 5 cc. is most often adequate. The anterior and lateral projection made

* Winthrop Laboratories Hypaque sodium is sodium 3,5-diacetamido-2,4,6-triiodobenzoate ($C_{11}H_5I_3N_2NaO4$) which contains 59 87% iodine.



FIGURE 3. Bilateral arteriography in child under general endotracheal anesthesia. Lead shield is placed between patient's head and site of injection in order to protect arteriographer's hands against radiation. A plastic band holds the patient's head in exact position in order to obtain true A.P. view without rotation.

with a seriographic apparatus is fairly well standardized. At times an oblique view is also made and in some clinics stereoscopic views are used. In case the procedure must be discontinued for any reason, the first films made should be those yielding the most information. The time allowed between injections is 10 minutes lessening the danger of complications, which fortunately are very few. The films are developed and read during this interval. Some surgeons prefer to do the bilateral carotid injections on a day separate from the vertebral injection while others perform this entire procedure at one sitting.

If it seems improbable that the vessel can be cannulated easily, French suggests a reflux brachial angiogram be done. This consists of direct visualization and cannulization of either right or left brachial artery in the mid portion of the upper arm. The basilar and carotid system are quite frequently adequately visualized especially if there is ipsilateral carotid artery compression (Figs. 7-8, Chapter XII).

The complaints following arteriography are relatively minor and are treated symptomatically with no restrictions.

Venography

Venography like arteriography is performed under general anesthesia. It is done by making a burr opening through the skull over the sagittal sinus just anterior to the coronal suture. (Before the anterior fontanelle is closed, it is not necessary to make the opening through the skull.) A 20 gauge polyethylene tube is inserted into the sinus. Gelfoam is placed over the top of it and the wound is closed. In the x-ray room, 4 to 8 cc. of Hypaque[k] are injected and seriographic films are taken in a manner similar to that described for arteriograms.

CHAPTER III

Electrodiagnostic Methods

CURTIS MARSHALL AND GEORGE AUSTIN

ELECTROENCEPHALOGRAPHY and electromyography, though not universally available, are described in this chapter in relation to their adaptability to the pediatric patient. Under electroencephalography consideration will be confined to trauma. For more details regarding its use in seizures, the reader is referred to the chapter on Epilepsy.

ELECTROENCEPHALOGRAPHY

CURTIS MARSHALL

An introduction to the electroencephalographic approach to brain injury in children cannot presume to instruct in the evaluation of such tracings. Thus, there is no place here for any exemplary illustrations of an insignificant few possibilities. Much more to the point is a presentation of the scope of the problem plus a brief outline of the types of electroencephalographic findings concerned and a second outline of the more likely applications of the methods.

Evaluation of the electroencephalograms of patients beneath the age of fourteen is a much more formidable task than with adult patients. The underlying cause for this is the comparatively tremendous breadth of the variability of the tracings seen in normal children. Existing studies of these normal patterns clearly demonstrate the mobile characteristics of the records observed in the changes with aging as well as the changes with varying stages of sleep and blood sugar level. The usual normal adult yardsticks of symmetry, clear frequency bracket, and stability with overbreathing mean little or nothing in the records of children. The resulting complexity of normality in children has brought many workers to the point of feeling that clear seizure discharges of either spikes or spike-and-dome are the only patterns which can diadactically be named "abnormal."

Unfortunately, all too few practicing physicians have access to the consultation services of an adequately trained electroencephalographer. In the United States, a majority of the EEG laboratories function with relatively inexperienced personnel both of medical and technical grades. This situa-

tion is not due to any inordinate demand for their services, but rather to the fact that so few doctors are aware of the years of training and experience necessary before the output of a laboratory can be relied upon. With this as a starting point, it would not be out of place to continue by bringing up *some items concerned with the adequacy and reliability* of an EEG laboratory.

Though the technique of producing a tracing in a child is essentially no different than with an adult, the following should be recognized:

(1) A multichannel (8 or more) machine is needed with a degree of similarity between channels beyond that which can be distinguished by a trained eye. Even a reliable machine requires constant checking on this point.

(2) Multiple electrodes (18 or more) should be used in an accurately reproducible pattern to cover the scalp areas over the cerebral cortex. Exact styles of electrode design are of little importance.

(3) Montages should be multiple with numerous (3 or more) cross checks of each scalp area. This is similar to viewing an object from three different directions in order to better appreciate its form.

(4) All records should be fully labeled as to time-constant, high frequency characteristics, paper speed and sensitivity. Also, the technician should note, on the tracing, special items such as clinical seizures, etc.

(5) Adequate activation procedures should be used comprising at least sleep and overbreathing when possible.

Reading of the tracings should be accomplished with the electroencephalographer having no knowledge of the patient other than his age. After the findings have been recorded, then the physician should consult the clinical record to seek a correlation between it and the electrical picture. Without such stringent regulation, it is all too easy for even the most conscientious electroencephalographer to be partly biased if he is familiar with the clinical picture before reading the tracing.

In taking advantage of the services of an electroencephalographic consultant, a physician must afford the time to furnish him with a complete account of the injury: time and date, direction of the blow, strength of the blow, patient's reaction in detail, both immediate and following, neurological examination, and sequelae. Also of importance are the patient's previous history as to birth injury, head injury, meningitides, personality variants, "febrile convulsions" and epilepsy. One must avoid such confusing terminology as "petit mal," "major motor," and "epileptic equivalent," as these and similar terms carry different meanings for each doctor. The previous history is of great importance in attempting to connect any EEG irregularities with their proper etiology.

Information of value in brain injury, then, falls under the following headings:

(1) *Changes with Time.* Pre-injury tracings are only available by chance. Thus, the main guide must be to obtain as early a post-injury tracing as possible. Then, serial tracings can show if there is a recovery curve. Three tracings should be obtained within the first month and probably others at about six months and one year.

(2) *Asymmetry of Voltage* alone in an otherwise normal tracing can be granted no significance under the age of one year. Beyond this age such asymmetries gradually gain significance but have only guarded significance at best.

(3) *Asymmetry of Rhythm* carries much more significance than that of voltage alone provided it is looked at with corrections for maturation. In many cases with sub-dural hematomata or cortical damage, the faster frequencies are less clearly seen over the involved hemisphere than over the normal one. On the other hand, the slower frequencies (less than 13/sec.) may show little or no asymmetry in the same case.

(4) *Slow Activity* cannot easily be labeled as abnormal in children unless there is a clear focus or a change with time. Bursts of slowing carry, at times, the connotation of abnormality provided there is no question that the patient is not dozing, a difficult thing to ascertain in a child. In addition, degree of electrical maturation varies widely for any given chronologic age and any slowing must be evaluated with these two factors in mind.

(5) *Seizure Discharges* are abnormal at any age and, when seen following injury, must be carefully documented with serial recordings and accurate localization technique.

(6) *Localization,* of all electrical evidence, is the most important. Thus, concise and flawless localization technique is mandatory. Every abnormal waveform must be localized and the area involved delimited.

(7) In all the above six items, the presence of a bony defect is a biasing factor and must be carefully evaluated in the electrical picture, be it of surgical origin or otherwise.

From the clinical point of view, there are certain aspects of injury in which EEG can play an important role:

(1) In suspected post-traumatic epilepsy, not only can the EEG differentiate with adequate certainty between idiopathic and symptomatic seizures, but it can also usually indicate the amount of brain tissue involved by the epileptic discharge and give some indication as to whether the area is cortical or sub-cortical. Thus, it is of aid in the consideration of possible surgical intervention.

(2) In the presence of a sub-dural hematoma, the EEG cannot make the diagnosis alone. However, if a sub-dural hematoma is suspected on clinical grounds, the EEG can usually point to the side or the area with reliability.

(3) In children, the EEG is a reliable tool for the investigation of a

possible brain abscess. The important point here is that the abscess itself is electrically silent while it is the normal brain tissue adjacent to the abscess which will generate the classic focal slow activity. Thus, the abscess can border on any aspect of the electrical focus.

(4) Gross brain damage can often escape the eye of the electroencephalographer in patients less than one year of age. However, whenever such an injury is suspected, repeat tracings during the second or third years are of value. Beyond the age of one year, minor brain damage can escape electrical notice but this does not negate the necessity of obtaining the tracing.

(5) In well documented concussion, EEGs on children often are negative. However, the same attitude as above maintains; i.e., follow-up EEGs are in order.

The electroencephalographer can, thus, be a valuable consultant in the handling of head injury cases provided he is not asked for a "blind reading." The electrical material can, in no manner, produce a diagnosis; it can only give a map of the cortical activity at a given moment. This information must be intelligently intercorrelated with the clinical information to aid in the final evaluation.

The use of EEG as an isolated measure of brain damage in law suits for personal injury is to be decried. Such prostitution of the technique is, nonetheless, often seen in courts.

Furthering of the application of EEG as an aid in head injury evaluation is under way in many clinical research centers and at the present time, government funds are financing a nationwide long term evaluation of cerebral palsy. Out of these studies will come a valuable amount of material to bolster the prognostic value of EEG in brain injury in children.

ELECTROMYOGRAPHY

George Austin

In spite of the value of a careful motor, sensory and reflex examination it is frequently important to enlist the aid of more quantitative electrical methods. These may be used as diagnostic aids, baseline determinations, and for prognosis. The problem of using these methods in children is made somewhat more difficult by the fact that they are uncomfortable and frequently involve the placement of fine needles through the skin. In spite of these minor objectives they present us with important data which should be available to anyone attempting the diagnosis and treatment of spinal cord or peripheral nerve pathology.

Historically these methods owe their original value to the pioneer work of Galvani, von Helmholtz, and Lapique. More recently the work of Kugelberg, Buchthal, Jasper, Marinacci, Grundfest, Hodes, and Eaton, has

added significantly to the literature on electromyography. It was in 1786 that Galvani first presented his evidence for so-called animal electricity, by showing that a frog muscle was capable of responding by twitch to a tiny electric current. The work of von Helmholtz (1850), a Prussian army surgeon, was unique in that he was the first to measure the actual speed of the nerve impulse (30 m./sec.), again using the frog nerve muscle preparation. Lapique, at the start of the twentieth century (1926), then showed that important diagnostic criteria could be obtained by measuring the values of rheobase and chronaxie. Utilization time as Keith Lucas called it had also been used similarly. Lapique was the first to show the practical value of these measurements as an estimate of excitability of nerves and muscle. Later it became apparent that the strength duration curve perhaps had more information to give than merely the measurement of chronaxie and rheobase.

With the application of the cathode ray oscilloscope to nerve physiology, the recording of multiple action potentials on volitional movement and in response to peripheral nerve stimulation, became known as electromyography. These were interpreted both as to frequency and pattern, and also in relation to the amplitude and shape of the individual action potentials. Both methods have their value and have been found useful clinically. Finally, the methods of direct neural stimulation for measuring conduction time, and the plotting of strength duration curves have been shown to have valuable applications. Just as direct neural stimulation has value, it has also been shown that direct muscle stimulation is also important in both determining the degree of degeneration and prognosticating the return of function by measuring the so-called Galvanic tetanus ratio.

Definitions

Rheobase: The least strength of current which if prolonged indefinitely will produce a threshold contraction of muscle.

Chronaxie: The minimal time that a current must flow, which is twice the strength of rheobase, to cause a threshold contraction of muscle. This is a measure of tissue excitability and must be used with care. Clinically the stimulating electrode is applied to the motor point and its value is therefor a combination of nerve and muscle chronaxie, although predominantly the former. The test should not be repeated more than once every ten seconds and should be compared with the opposite and normal muscle if possible. (See Fig. 1.)

Strength-Duration Curves: A graph of the strength of stimulus necessary for threshold movement, plotted against the time the stimulating current must flow on the abscissa. Actually this is a more sensitive index than

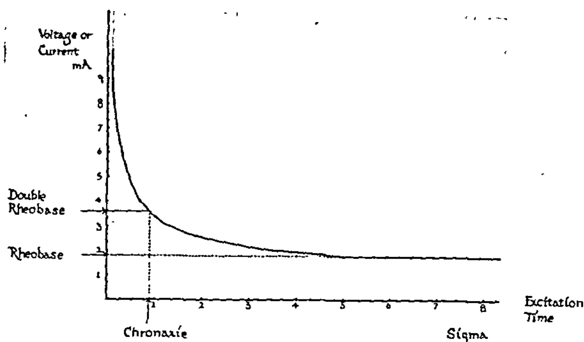


FIGURE 1. Normal form of strength duration curve to illustrate excitation time, rheobase, and chronaxie.

chronaxie, which is merely one point on the curve. Hence the curve may be shifted up or down without change in chronaxie, yet with marked difference in tissue excitability. (See Figs. 2 and 3.)

Tetanus Ratio: The ratio of the Galvanic current required for a sustained tetanic contraction to that current required for a threshold contraction.

Threshold: When applied to muscle it is that twitch which is just apparent to the observer.

Latency: The time between the stimulus artifact and the start of the action potential, as seen on the cathode ray tube.

Conduction Time: Synonymous with latency. Used in measuring the velocity of nerve impulse, by stimulating two points on the nerve and then measuring the difference in latency. Dividing this into the distance between stimulating electrodes gives the velocity of conduction and was the method originally used by von Helmholtz in both frog and human.

Fibrillation Potential: The electrical activity of a single fibril, thought by some to be hypersensitivity to a chemical agent in the muscle. A sign of a completely denervated fiber. Usually about 100 micro volts amplitude and less than a half millisecond in duration. (See Fig. 4.)

Fasciculation: Contraction of a muscle fascicle which is usually visible to the eye of the observer. Increased by cold or tapping the muscle. Can occur as a single potential of several hundred micro volts and about one millisecond long, but usually present in bursts of activity. (See Fig. 5)

Fatigue: A normally occurring phenomenon in muscle consisting of a progressive decrease in the rate of firing of motor units without change in amplitude. Pathologically, as in myasthenia gravis, there is an associated decrease in the amplitude of the individual units as they decrease in frequency of firing. (See Fig. 6.)

Motor Unit: Consists of the anterior horn cell, its axon, and the number of muscle fibers supplied by the axon, usually approximately 200 fibers. Normally fires 5-10 times per second on volitional movement and as the strength of stimulus is increased additional motor units begin to discharge.

Insertion Potential: That series of potentials which follows insertion of the recording needle into the quiescent muscle. Normally lasts a second or less and consists of a series of normal and polyphasic action potentials. In myotonia it is exaggerated and consists of a prolonged train of spikes and positive waves. Is also increased in denervated muscle and polymyositis. Severe disintegration of muscle fibers causes reduced insertion activity.

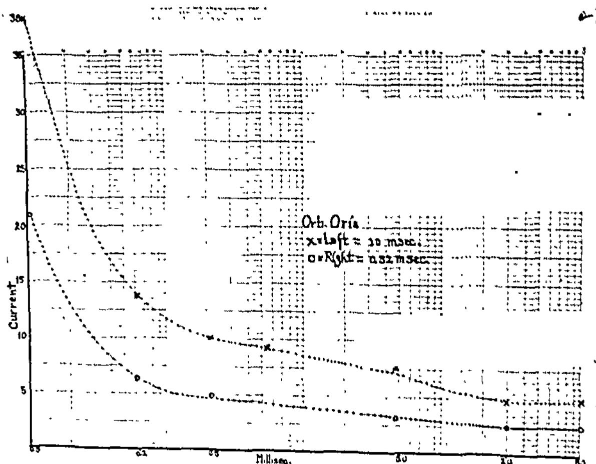


FIGURE 2. Strength duration curve plotted in case of peripheral facial nerve injury on the left. The strength duration curve depicts the difference between the two sides more accurately than the chronaxie values.

Practical Evaluation

In determining the usefulness of the various methods and trying to evaluate their importance to decide whether or not a test should be used, one should keep firmly in mind what information can be obtained by the use of so-called electro diagnostic methods. The following are practical factors to consider.

Differentiating an Upper Motoneuron from a Lower Motoneuron Lesion

One may often obtain clinical evidence by signs of visible atrophy or fasciculations that there is a lower motoneuron lesion present, and further corroborative evidence may be obtained by seeing whether the reflexes are hyper or hypoactive. Occasionally there may be a difficult point where one cannot be sure whether one is dealing with an early lower motoneuron paralysis or an actual upper motoneuron lesion. In these borderline cases, determination of strength duration curves and the measurement of chronaxie may be of extreme value. This is probably one of the most delicate means of detecting early lower motoneuron damage; i.e., by a change

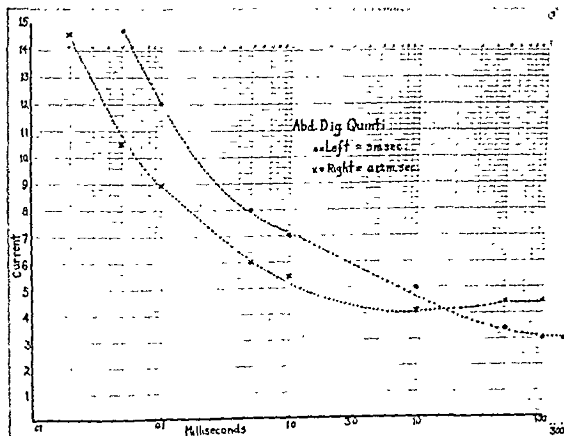


FIGURE 3. Strength duration curve in a case of ruptured cervical disc at the C6-C7 level on the left.

in the shape of the early portion of the strength duration curve or an increase in chronaxie time.

Electromyography may also be important here. The finding of fasciculation potentials, a decreased number of motor units, or increased amplitude of individual motor units are all evidence of a lower motoneuron lesion.

Differential Diagnosis Between Root, Nerve, or Muscle Disease

When a compressive lesion involves one or more roots, there will be corresponding changes in excitability in several nerves without complete signs of degeneration in any one. This, of course, can be found most clearly in cases of spinal cord tumor with segmental root compression at the level of the tumor. These changes of excitability may be clearly seen by measuring the chronaxie values of those muscles displaying weakness or other signs of dysfunction. Ordinarily such values would show signs of elevation in the range of 2-6 milliseconds. Electromyography would show fewer motor units firing in the involved muscle in cases of root compression. In addition, occasional fibrillation and fasciculation potentials may be found in patchy areas in the muscle.

In contrast to this, cases of peripheral nerve involvement will show more signs of complete degeneration. In these muscles chronaxie value will usually be greater than 10 milliseconds and there will be a great paucity of normal motor units firing, depending on degree of involvement. Fibrillation and fasciculation potentials will be more common and the muscles involved will be few and confined to a single peripheral nerve distribution. The galvanic tetanus ratio will be decreased. Action potentials will be smaller and the conduction time prolonged when compared with the opposite normal side.

When dealing with diseases of the myoneural junction or primary muscle involvement a somewhat different type of response is obtained from the electromyogram. Excessive fatigue in response to a repetitive stimuli will be found in such diseases as myasthenia gravis. Hence, motor units will be found to decline in amplitude as well as frequency, although this same picture may occasionally be seen also in poliomyelitis and amyotrophic lateral sclerosis. Also when there is primary muscle disease the motor unit action potentials will be smaller in amplitude and a relative increase in frequency will be found per equivalent contraction.

On needle insertion in primary muscle disease one may encounter a myotonic discharge with multiple positive waves of decreasing amplitude.

To Estimate the Degree of Peripheral Nerve Injury

Chronaxie values will give an approximation by the amount of their increase over the normal side. Similarly the reduction in the tetanus ratio

is helpful in estimating degree of injury. Electromyographic studies are important here in showing the number of normal motor units and number of excessively large motor unit potentials per unit of time. Likewise, the frequency of motor units will be progressively decreased as peripheral nerve damage increases. Fibrillation potentials are usually found only with complete denervation. Fasciculation potentials occur frequently and early in peripheral nerve lesions although previously it was thought that they were purely a sign of anterior horn cell disease.

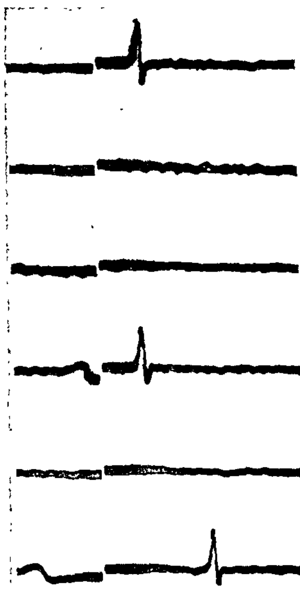


FIGURE 4 Fibrillation potentials in case of lower motoneuron damage Amplitude approximately 100 microvolts and duration 0.5 millisec. Note that the baseline is completely silent except for the fibrillation potentials

To Measure Return of Function and Analyze the Source of Poor Return of Function

It has been found that chronaxie values after a peripheral nerve section rarely return toward normal before there is some evidence of functional return. However, there may be earlier changes in the strength duration curve and therefor it is important to follow this at intervals during the proposed course of regeneration following a peripheral nerve lesion.

Voluntary movement of muscles may be impaired for any of the following reasons following a peripheral nerve lesion.

(a) Failure of sufficient fiber regeneration within the nerve.

(b) Lack of innervation of the muscle.

(c) Absence of normal muscle fibers due to excessive atrophy or fibrosis.

(d) Failure of relearning to use a muscle after injury to the nerve.

(e) Hysteria or malingering

Volitional attempts to move a muscle may prove unsuccessful, yet direct neural stimulation above the lesion may cause the muscle to function. Under these circumstances one is forced to the conclusion that the patient is malingering, hysterical, or else has failed to relearn functional use of the reinnervated muscle. Under the latter conditions it may be that the central pattern of learning has been rearranged. However, self stimulation and teaching the patient to reuse the muscle by showing him how it works are usually sufficient to gradually bring about functional return. Malingering and hysteria may be more difficult problems, although usually in hysteria there will be bizarre sensory changes and other side factors to support the diagnosis.

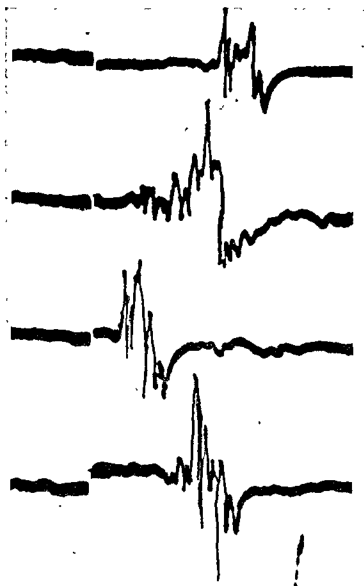


FIGURE 5 Fasciculations of biceps muscle in case of lower motoneuron disease as they appear on cathode ray tube. Amplitude approximately 0.5 millivolts and duration approximately 10 millsec.

Insufficient fiber regeneration for movement may usually be validated by the electromyograph. Chronaxie studies will show an elevation due to the large population sampled. There will be a paucity of motor units and many of increased amplitude under these conditions. In addition, there may still be some fasciculation potentials present. If the muscle is insufficiently innervated for voluntary contraction there will be a much slower contraction than normal with direct stimulation, as during the chronaxie studies. Direct nerve stimulation above the lesion may fail to produce a contraction, yet stimulation below the lesion produces a contraction. This can only occur if insufficient fibers are present across the site of the lesion, or when there is an early nerve lesion with a so-called propagation block. In the latter case, however, the chronaxie value would be relatively normal.

The tetanus ratio begins to rise as regeneration of a peripheral nerve occurs and then gradually falls to normal limits. Failure of innervation of a muscle may be determined by direct neural stimulation. In this case no muscle action potential is recorded or else an abortive and prolonged action potential is found.

Methodology

1. Direct Nerve Stimulation

(a) Stimulate through the overlying skin or by direct intraneural stimulation.

(b) May use monopolar or bipolar electrodes.

(c) In peripheral nerve injury, stimulate above and below the block or site of injury.

(d) Observe the type of muscle twitch and note the threshold.

(e) Record the muscle action potential to obtain latency.

(f) Measure speed of conduction of nerve impulse, if possible, to stimulate at two different points on the nerve.

(g) Whenever possible compare the normal with the abnormal side.

(h) Test the response to single and repetitive stimuli to measure fatigue.

2. Rheobase, Chronaxie, and Strength-Duration Curves

(a) Use either rectangular wave pulses of variable pulse duration, or condenser discharges of variable capacitance.

(b) Electrodes should be monopolar and of sufficient size to cover a satisfactory area of muscle (3-10 mm.) when stimulating over the motor point.

(c) Plot time on abscissa and strength of stimulus on the ordinate when using strength-duration curves.

(d) For chronaxie use same size electrodes and compare normal with abnormal side whenever possible. Normal is less than 1 m. sec. duration.

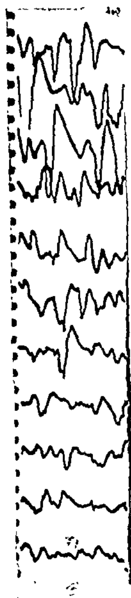
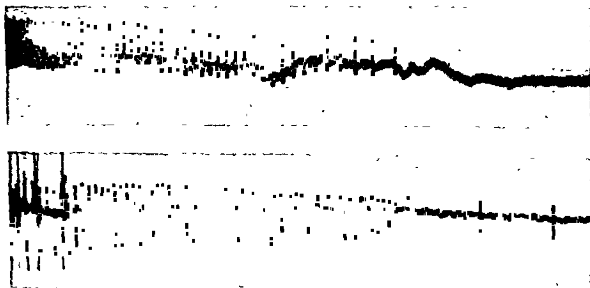


FIGURE 6. a. Shows fatigue on voluntary contraction of opponens pollices muscle in case of myasthenia gravis. Successive sweeps are 5 sec. apart. Muscle potentials are 2-2.5 millivolts maximum. Duration of each sweep = 30 milliseconds.

b. Fatigue of motor units on voluntary contraction of opponens in case of myotonia. Note decrease in amplitude as well as frequency. Length of sweep = 110 millisec.

c. Fatigue of motor units in case of myotonia. (Gastrocnemius) Length of sweep = 150 millisec.



(e) Strength of stimulus may be recorded in volts or milliamperes. Theoretically a constant current stimulator with output maintained constant in spite of resistance changes, is best.

(f) If time permits, a strength-duration curve is better than merely obtaining rheobase and chronaxie values.

(g) Compare with opposite side if possible.

3. *Galvanic Tetanus Ratio*

(a) Use a galvanic current, preferably calibrated in milliamperes output.

(b) Observe muscle for twitch and record this threshold strength of stimulus.

(c) Observe muscle for sustained contraction (Tetanus) and record this strength of stimulus. This should be done for a contraction of at least four seconds duration.

(d) Record the ratio of tetanus to twitch currents. Normally this is 3 to 5.

4. *Electromyography*

(a) May use monopolar or bipolar recording. Monopolar is best done with an insulated hypodermic needle and the larger indifferent electrode on nearby inactive tissue such as skin or tendon. Bipolar is best done with concentric needle electrodes with an insulated wire cemented within a hypodermic needle shielded down to the tip.

(b) Observe and record important or characteristic responses by photographing the cathode ray tube response. Use a high gain, differential input, preamplifier.

(c) Observe the response to needle insertion, volitional contraction, and nerve stimulation.

(d) Look for fibrillation potentials and fasciculation potentials with the muscle at rest, when denervation is suspected. The former are due to single fibrils contracting. The latter may be single but more frequently occur in bursts.

(e) Look for signs of excessive fatigue during volitional contraction as well as abnormal amplitude and frequency.

(f) Finally increase the sweep speed and examine the potential shape in abnormal muscle. The amplitude is greater and the duration prolonged, with more polyphasic responses in polio and in reinervating muscle.

(g) Again, compare the normal with the suspected abnormal side whenever possible.

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CHAPTER IV

Principles of Pre- and Postoperative Care

THEODORE C. PANOS

IT HAS PROPERLY been emphasized that progress in understanding of nutrition, particularly of fluid and electrolyte metabolism, has been a primary factor in the development of the spectacular achievements and safety of modern surgery. In the case of infants and children, there are several reasons for devoting special consideration to the circumstances surrounding surgery. *Homeostatic* responses which are ordinarily protective *may be immature or incompletely developed, e.g., exaggerated tendency* of newborns and small infants to retain sodium and water and their relative inability to produce concentrated urine, with the resultant contrasting dangers of water and solute loading and of dehydration. The rapid changes induced by continuous growth and development create quantitative and qualitative nutritional requirements which are different from those in the adult. Special precautions are necessary to preserve emotional stability and development.

Whereas these remarks apply to all forms of surgery in this age group, they are particularly pertinent to neurosurgery. Because most procedures in this field are of considerable magnitude and duration, the stresses are more profound and thus more challenging to processes of homeostasis and repair. Because of the varying and complex role of the central nervous system in the control of electrolyte and water balance, disturbances induced by surgery may result in the following changes, any one of which could be dangerous: retention of water and sodium; simple water retention; salt diuresis; or simple water diuresis. Proximity to centers controlling a variety of homeostatic mechanisms increases hazards of such acute complications as shock, coma, respiratory obstruction or failure, and infection. When all these considerations are kept in mind, it can be said that infants and children tolerate the wide range of neurosurgical procedures surprisingly well. In view of remarkable technical advances, all techniques and special procedures applicable to adults are also applicable to infants and children.

Success, however, depends on meticulous attention to details of observation and management. The accurate assessment and provision of pre- and postoperative fluid and electrolyte needs are essential to successful surgery.

Preoperative Measures

Clinical observation, based on thorough history and physical examination, will dictate the approach to the newly admitted neurosurgical patient. Frequently, there has been no occasion for development of significant deficit in fluid, electrolyte or calories. The usual routine studies are obtained. Baseline and daily body weights should be carefully recorded. Regular diet is provided, with emphasis on fluid and carbohydrate intake. Because body stores of carbohydrate and fluid are quickly exhausted, the period of starvation prior to surgery should be as brief as possible, e.g., 6-8 hours in an infant. Whenever possible, pediatric cases should be scheduled first during the day, in order to avoid prolongation of starvation, as well as to ensure that a venous cut-down will not have been done in vain, in the event the surgery is cancelled due to difficulties with the previous case. Blood transfusion before operation is indicated only when hemoglobin is 10 gm.% or below (after rehydration), since it is wise to avoid the hazard of hypervolemia with its possible complications on the occasion of administration of blood during surgery, which is essentially routine. Cathartics, especially saline preparations, are generally to be avoided, although, as will be mentioned, fecal impaction must be guarded against in postoperative state. Gastric lavage to empty contents is advisable in comatose patients, prior to surgery.

Because neurosurgical procedures are frequently extensive and very stressful, it is imperative in the vast majority of cases that an intravenous infusion be securely established before surgery is undertaken, preferably by "cut-down" in the usual case. Such a measure yields triple assurance: it will enable replacement of losses during surgery; it will enable prompt and energetic therapy of shock at the auspicious moment of need; and it will provide for the variable and often extended period of parenteral feeding postoperatively. At the time of cannulation, it is wise to add a three way stopcock to the system in order to allow for special rapid infusions, e.g., hydrocortisone and blood in the event of shock.

If vomiting has been conspicuous and persistent prior to admission, combined fluid and electrolyte (chiefly chloride and potassium) deficits will be present as well as evidence of caloric undernutrition. In such an instance, for example, the administration of dextrose in distilled water would seriously exaggerate hypoelectrolytemia, whereas administration of saline solutions would greatly aggravate potassium loss and contribute to the possibility of sodium overloading. Additional initial studies are, there-

fore, frequently very helpful, if not necessary, for guiding preparation for surgery. In addition to serum carbon dioxide content, chloride, sodium and potassium, the determination of non-protein nitrogen, hematocrit, and total protein may be especially helpful in evaluating the patient and in establishing baseline observations for reference during what is commonly a stormy postoperative course. The availability of microtechniques greatly facilitates these measurements, and consequently reduces the threshold of indication. Blood transfusion before surgery is often necessary. Attempts to restore positive nitrogen balance by energetic feedings are, however, usually not feasible, since the urgency of surgery outweighs this consideration, especially when it is possible to make full water and electrolyte restitution before surgery.

Whenever possible and feasible, preoperative sedation should be so managed that the patient is asleep or at least oblivious of surroundings by the time of arrival to the operating suite.

Of particular importance is the proper psychologic preparation of the patient and family. Children, by virtue of their immaturity and imaginative nature, are unusually susceptible to anxiety and fear reactions resulting from the many formidable and new experiences in a hospital environment and from separation from parents. A minimum of explanation is frequently better than too much, since the capacity for misunderstanding exceeds that for true understanding, which in itself is very limited. Reassurance and emotional support in the form of affectionate personnel and maximum parental attendance will more than compensate for failure to provide detailed explanations. Also, it must be recalled that parental reaction to the illness and projected surgery, if not sound, can result in much unnecessary and harmful apprehension and insecurity in the child. It is, therefore, imperative that the neurosurgeon devote careful attention to frank discussions with parents and other directly involved persons during all phases of the relationship.

Care During Surgery

The gravity of most neurosurgical procedures in this age group and the difficulty of anesthesia dictate the desirability of the constant presence of a "floating" physician whose chief duties would be to supervise the administration of fluid, blood, and medications and to assist the anesthetist, particularly in the event of shock. The rectal temperature should be carefully followed in order to prevent overheating and wide fluctuations in temperature. Blood transfusion during the procedure is almost routine in amounts depending on loss and the degree and duration of trauma. It is unwise to begin surgery without the certainty of availability of blood in the operating suite. Generally, the total ... given ... being

estimated loss plus 20-30 ml./kg. The amount of saline used to precede and follow the blood should be rigidly minimized because of the well known tendency and danger of retention of saline and water, particularly in infants. Plasma should be avoided if possible, in view of the danger of hepatitis. Solutions containing potassium should never be administered during surgery, because of the amount of the ion liberated during trauma and the danger of high levels in shock or "shock-phase." The ready availability of such intravenous preparations as hydrocortisone, norepinephrine and dextran and of a "cut-down" tray should be assured before the procedure begins.

Postoperative Management

Attention to detail, according to an organized plan of observation and evaluation, constitutes the only promise of uncomplicated postoperative recovery. The establishment and routine use of the "recovery room" has provided indispensable specialized nursing care and facilities for early detection and prevention of acute complications such as shock, respiratory obstruction, aspiration of vomitus and secretions, and urinary retention. This is of particular importance with pediatric patients in whom changes may be more labile and difficult to discern or interpret for personnel not specifically indoctrinated for this purpose.

Of equal importance, as clearly indicated by numerous extensive studies in recent years, is full realization that metabolic changes induced by surgery are profound and may extend over a prolonged period of days or even weeks. Failure to cope specifically with these changes can be fatal, and, in the past, was a major cause of discouraging morbidity and mortality rates. Although there are considerable individual variations, the pattern of response is similar in all cases, regardless of the nature of the surgery, which may, however, exaggerate certain aspects of response or exert special effects. With the probable exception of newborn infants, qualitative changes do not differ significantly from one age group to another. Discussion of these changes, in addition to those of mechanical or infectious origin, can perhaps best be handled by considering each potential hazard involved more or less in order of chronology and importance.

The danger of *laryngeal obstruction* is so great in certain circumstances that prophylactic tracheotomy in the operating room has achieved ever widening usage. Aside from neurologic factors and tendency for laryngospasm, the prolonged endotracheal intubation required for safe and effective anesthesia, the small laryngeal diameter and the awkward positioning of many patients (trunk erect with head flexed) all predispose to laryngeal edema and obstruction. The smaller the patient, the more respiratory

obstruction is to be feared. In all infants and children, the provision of a highly moisturized atmosphere during the early postoperative period is important. This is best accomplished in an incubator or "croup" chamber (Croupette®); most observers agree that the addition of such preparations as Alevaire® is not significantly beneficial.

In this connection, it is also of importance to keep the patient on the abdomen as much as possible, in order to take advantage of the anatomic drainage gradient of the respiratory tract. The tendency for hypoventilation plus that for aspiration make the supine posture a hazardous one, not to mention elevation of the head and shoulders. Because the latter position is often necessary after surgery, the patient should be particularly closely observed for respiratory complications.

There are certain well established changes in fluid and electrolyte physiology following surgery. Retention of sodium, increased urinary excretion of potassium (and usually chloride) are conspicuous, and may last for 2-5 days or even longer. Retention of water accompanies that of sodium, and there is a consequent tendency for edema. These changes have all been ascribed to increased production of aldosterone by the adrenal cortex and the clinical result is the tendency toward postoperative alkalosis with hypokaliemia and hypochloremia. The loss of potassium is aggravated by administration of sodium-containing fluids. The familiar injunction against injudicious use of saline, especially during surgery and the first postoperative day, is thus well founded and should particularly be observed in newborn and young infants. A commonly employed routine is to administer no sodium-containing fluid during the first postoperative day, one-half the calculated requirement on the second, and the full requirement on the third. Likewise, it has also been found wise to provide only two-thirds of the calculated fluid requirement during the first and possibly the second postoperative day.

On the other hand, there are several considerations which emphasize the dangers of *hyponatremia* ("low-sodium syndrome").

There is a strong antidiuresis, presumably of hypothalamic origin, especially early after surgery (18-36 hours), leading to water retention unaccompanied by sodium retention. This reaction may predominate over the usual aldosterone effect, and in the course of providing intravenous fluids without sodium, the administration of an excess of fluid predisposes to water intoxication.

Lesions of the central nervous system, including surgical, are frequently associated with increased renal excretion of sodium (and chloride).

Overloading with intravenous solution of dextrose in distilled water occurs frequently due to error in calculating fluid requirement, too rapid administration and use of large fluid containers which can easily deliver an overload to a small patient.

Occasionally, dextrose in distilled water is very ill advisedly given subcutaneously in the postoperative period, with resultant rapid withdrawal of electrolyte from circulation into the area of hypodermoclysis. Serious depletion of sodium may thus occur and markedly predispose to water intoxication.

Preoperative losses due to vomiting, diarrhea, and perspiration will exaggerate electrolyte deficits and tendency to water intoxication.

Awareness of the clinical manifestations of water intoxication is of crucial importance, because they may simulate neurologic changes associated with the primary disorder and/or the neurosurgery. Restlessness, irritability, confusion, headache, vomiting, convulsions and stupor or coma may occur, as well as papilledema and retinal hemorrhages. When faced with such a picture in a postoperative neurosurgical patient, it is obvious that the immediate determination of serum electrolytes is indicated, as well as an assessment of all aspects of preoperative, surgical, and postoperative management which might have led to electrolyte (Na) deficit and/or water excess.

Potassium ion has assumed particular importance in postoperative care because of the serious and potentially fatal consequences of deficit or excess (particularly the former) and the ease with which these derangements can be produced. Renal excretion is greatly increased for two to five days or longer following surgery, concomitantly with increased retention of sodium and water. Administration of potassium-free solutions, especially saline, exaggerates renal loss of this cation. Deficiency may also result from recurrent vomiting, gastric suction, diuresis from any cause, and diarrhea. It should be emphasized that no body fluid contains less potassium than plasma. Important clinical manifestations are anorexia, vomiting, weakness, abdominal distention, tachycardia with cardiac dilatation, and even myocardial decompensation. There is a tendency for hypochloremic alkalotic tetany, especially in infants, which cannot be corrected without addition of potassium salts.

Parenteral potassium need not be given for twenty-four to forty-eight hours postoperatively because of the amount available from tissue breakdown at surgery. Thereafter, supplementation is necessary in patients receiving nothing by mouth, in amounts of about 3 mEq/Kg./24 hr. (or more if there are continuing losses via suction, vomiting, etc.). The concentration of fluid containing potassium should not exceed 40 mEq/liter and administration should be spread out over the twenty-four hours. Patients receiving potassium intravenously should be carefully observed for signs of excess (bradycardia, irregular rhythm, peripheral vascular collapse, apprehension and weakness). Potassium should not be given if there is any evidence of impaired renal function. As soon as oral feedings can be tolerated, potassium requirements can easily be provided.

Excessive drainage of cerebrospinal fluid from wounds, etc., should be replaced volume for volume with physiologic saline in the calculated daily oral or parenteral fluid requirement.

Adynamic ileus may occur after any operation. In addition, the role of potassium deficiency in the production of ileus is well known. Oral feedings should not be begun until return of peristaltic function has been assured.

Maintenance of nutrition may be easily neglected in over-all postoperative care. Protein metabolism, being of particular importance in wound healing and resistance to infection, is greatly disturbed following surgery. Nitrogen excretion is greatly increased after all operations, especially after extensive procedures. This "catabolic response to injury" lasts usually several days (two to seven days). Preoperative starvation and inadequate intake postoperatively aggravate the depletion of protein stores and adversely affect recovery. Patients should be fed as soon as possible and returned to a full diet as quickly as possible. Feeding by gastric tube should be freely employed. The progression should be from clear fluids to surgical liquids, skim milk, whole milk, soft diet, and full diet. Vitamin supplementation is important, especially vitamin C, but B complex should also be provided, Vitamin K is often advisable in newborn and small infants.

General Remarks

Estimation of fluid and electrolyte requirements should be based on evaluation of as many pertinent observations as possible: history and physical examination, body weight, temperature, intake and output, urinalysis, hematocrit, etc. Certain basic measurements as *body weight* and urine volume and specific gravity are especially useful for assessing day-to-day variations in hydration. It should be recalled, however, that adequate to large volumes of dilute urine can occur in sodium-depleted patients receiving water by mouth or glucose in water parenterally. Repeated determination of electrolytes in serum and discharges may be necessary to guide therapy during parenteral maintenance. Indeed, serum electrolytes should be routinely studied at twenty-four and seventy-two hours following neurosurgery, and every forty-eight hours thereafter as indicated, in order to detect dangerous fluctuations in sodium (sodium and water retention, sodium diuresis and/or simple water retention) and potassium (especially hypokaliemia). The serum sodium concentration can be approximated (in the absence of ketosis) by adding fifteen to the sum of carbon dioxide content and chloride concentration. Serial electrocardiograms are helpful in reflecting abnormalities in serum potassium concentration.

Requirements may be expressed as milliliters of water or milliequivalents of electrolyte per kilogram of body weight, per square meter of body

surface or per 100 calories metabolized. Calculation according to weight is conventional and particularly useful in dealing with fluid and electrolyte problems. Calculation according to surface area represents a simpler and increasingly popular clinical practice. The following table lists approximate daily requirements in terms of both weight and surface area:

Age-Years	*Ml./Kg.*	*Ml/M²*
Infancy	150	1500
1-3	125	1500
4-6	100	1500
7-9	75	1500
10-12	75	1500
13-15	50	1500

Sodium: 2 mEq/Kg. or 40-60 mEq/M²
Potassium: 3 mEq/Kg. or 40-60 mEq/M²

It should be emphasized that special problems attend the care of newborn infants undergoing surgery. Caloric and fluid requirements are much less than in older infants and children. Because of renal limitations, sodium tolerance is especially poor. Overhydration and edema are thus important hazards to avoid. Also, it has been found that newborn infants probably do not respond to surgery with the usual exaggerated urinary loss of potassium observed in older individuals. On the basis of these considerations, the following may be listed as postoperative requirements in newborn infants:

Water:	40-50 ml./Kg. for the first two days
	50-100 ml./Kg. daily thereafter
Sodium:	nil for first 2-3 days
	1 mEq/Kg daily thereafter
Potassium:	nil for first 2-3 days
	2 mEq/Kg. daily thereafter

Intake and output should be carefully recorded and checked. Discrepancies between what is ordered and what the patient received may be serious. Exact knowledge of fluid and electrolyte intake is important and relatively easily accomplished with good nursing assistance. Collection of gastrointestinal discharges, particularly gastric, is of great importance in enabling ordering of accurate replacement. Although analysis of electrolyte content of these discharges represents the ideal basis for replacement calculations, the following table lists figures for approximate compositions which can be reliably used in estimating specific replacements. Actually, it is common practice to replace gastric losses volume for volume with isotonic saline plus at least 10 mEq of potassium per liter.

	Gastric Secretions	*Intestinal Secretions*
Sodium	30-60 mEq/l	100-140 mEq/l
Chloride	100-150 mEq/l	80-120 mEq/l
Potassium	10-20 mEq/l	10 mEq/l

ing. The proper combination of antibiotics used to prevent postoperative complications will, therefore, vary according to the incidence, and sensitivities of infections observed in a particular hospital over a period of time. The duration of such therapy should be as brief as feasible in order to discourage increasing the resistance of such organisms.*

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* Since this chapter was written new antibiotics have entered the field Albamycin is now, December, 1958, being used for Phage 81 and in resistive cases, Kanmycin and other new antibiotics have been successful. Antibiotic resistant bacteria will continue to be a serious problem which will be conquered only when antibiotics are no longer routinely administered for non-specific infections and illnesses —I. J. J, *Editor*

CHAPTER V

Anesthesia

Charles R. Allen and Leo S. Duflot

THE EXTREMELY narrow margin of error permissible for neurosurgical procedures in children imposes continued psychic stress upon the neurosurgeon and emphasizes the desirability of freeing the operator from distracting considerations concerning the general condition of the patient. The physician anesthetist should assume the duties of clinical physiologist in the operating room. He should be cognizant of the pathological physiology of the patient's disease and its associated complications as well as the technical details of the operation itself. The anesthesiologist's challenge should be to maintain the patient in as near a normal physiological condition as possible. Small variations in clinical signs may indicate beginning stress from blood loss, fluid imbalance, hypoxia, hypercarbia or "retractor" ischemia. The early diagnosis and correction of these conditions may be a vital factor in the survival of the patient. In order to accomplish these things it is mandatory that the pulse rate, blood pressure, respiration and other available indicators be followed, recorded and evaluated throughout the procedure.

For satisfactory management the following circumstances should prevail: (1) Depression from premedicant drugs should be avoided. (2) Induction of anesthesia should be smooth. (3) Resistance to inhalation or to exhalation should be at a minimum. (4) Light levels of anesthesia should be maintained. (5) Positions which allow embarrassment of respiratory or circulatory functions should be avoided. (6) Intracranial pressure should not rise above the preoperative level. (7) The surgeon should have free access to the operative site. (8) Blood, electrolyte and water replacement should proceed according to the estimated loss. (9) Emergence from anesthesia should be rapid.

The patient's pre-existing disease and physical status may increase the difficulty of maintaining these desirable circumstances.

Preoperative Preparation

Atropine or scopolomine should be used to suppress secretions during anesthesia. The dosage varies between 0.08 mg. and 0.32 mg. and should

be given thirty to ninety minutes before induction of anesthesia. In the presence of increased intracranial pressure, as with tumors involving the brain stem, even small doses of morphine, meperidine or a barbiturate may produce an exaggerated depressant effect upon respiration and may also mask vital neurological signs. For this reason they should not be used in patients scheduled for intracranial procedures. For neurosurgical procedures not involving the head morphine or meperidine may be included in the premedication to facilitate satisfactory anesthetic management.

An empty stomach should be assured prior to induction. In elective procedures this may be accomplished by the limitation of oral foods and fluids; however, water and electrolytes should be adequately maintained by intravenous infusions. In emergency procedures the stimulation of the gag reflex may induce vomiting. The dangers from an increased intracranial pressure must be carefully considered and may contraindicate the utilization of this maneuver. After the trachea is intubated the stomach may then be safely evacuated by inserting a tube.

If a marked increase in intracranial pressure secondary to the neurological disease is evident, a smooth induction will more likely be accomplished if the pressure is released by ventricular puncture prior to the beginning of anesthesia.

Choice of Technique and Agents

The technique of choice for all children and infants, except those who are comatose and areflexic, is general inhalation anesthesia. Inhalation agents are not metabolized, and they may be rapidly eliminated if the child's general physical condition should begin to deteriorate. Some clinics have preferred local and topical anesthesia with restraints because of their experience with unsatisfactory operating conditions associated with general anesthetics. Critical studies by Stephen indicate that it has not been the anesthetic agents but rather the techniques with which they were being administered that resulted in unfavorable experiences. If the narrow permissible physiological "margin of error" is not exceeded, infants and children will tolerate general anesthesia for surgical procedures of eight hours or more and may react promptly at the conclusion of the surgical procedure.

Children who are comatose may not require a general anesthetic at the beginning of the surgical procedure. They do require the services of a physician experienced in the maintenance of an open airway, tracheobronchial aspiration, adequate ventilation, and blood and fluid replacement therapy. If the operation improves the patient's physiological condition a general anesthetic may become necessary to properly conclude surgical procedure.

The choice of agents should be left to the physician anesthetist. A com-

bination of drugs which proves highly satisfactory when used by one anesthetist may fail miserably when employed by an individual inexperienced in their use.

Induction

In patients requiring intracranial procedures the period of induction is one of the most critical. Voluntary breath-holding, laryngospasm and apnea due to profound anesthesia should be carefully avoided. Induction with nitrous oxide and oxygen is preferred because of its non-stimulating properties. It can be used to introduce other agents such as cyclopropane or ether until anesthesia with the more potent drug is well established. Sodium thiopental can be used as an inducing agent in healthy older children with no preoperative respiratory or circulatory depression. It should be used carefully and sparingly. The rapid injection of even small amounts of this drug may be dangerous to the child with increased intracranial pressure.

Intubation

Endotracheal anesthesia not only provides for the administration of oxygen, exclusion of blood and mucus from the lungs, and the control of anesthesia but plays a major part in our efforts to help control blood loss and prevent further increase in intracranial pressure.

The effects of straining against a closed glottis during a stormy induction has resulted in measured intracranial tension of nearly 2000 mm. water, nearly twice as high as occurred subsequently in the same patient during "bucking" on an endotracheal tube.

There is no age limit which contraindicates intubation of the trachea. This procedure should be done in most children to facilitate the maintenance of an open airway throughout the surgical procedure. To avoid trauma to the glottic area, as well as coughing, bucking and laryngospasm anesthesia should be well established before intubation is attempted.

The anesthesiologist should have available at his fingertips a suction catheter, a laryngoscope of proper size and in good working order and various sizes of oropharyngeal and nasopharyngeal airways. He should also have within reach several endotracheal tubes of various lengths and bores larger and smaller than the one he has chosen to insert.

When anesthesia is of sufficient depth to relax the jaw and obtund laryngeal reflexes intubation may be attempted. A pillow to support the occiput is desirable, particularly in older children. In most cases this support provides for a more direct anatomical approach to the epiglottis. It is also helpful in preventing extreme extension of the neck, an undesirable situation in traumatic conditions of the cervical spine or where herniation of the brain stem is imminent. The patient's mouth is opened

widely and the laryngoscope blade is introduced along the side of the tongue. The epiglottis is gently lifted to expose the vocal cords. The tip of the endotracheal tube is quickly introduced so that it lies approximately two or three centimeters beyond the cords. A bite block is placed between the teeth and the source of anesthetic gases is immediately attached to the proximal end of the tube.

It is often desirable to apply a topical anesthetic to the mucous membranes of the pharynx, glottis and trachea. This act itself may stimulate severe laryngospasm if general anesthesia has not been well established. However, in deeply comatose children general anesthesia may be avoided entirely if this procedure is carried out before the trachea is intubated. Tracheobronchial toilet may subsequently be performed without reflex stimulation.

If the patient's general condition does not permit of sufficiently deep anesthesia to relax the jaw and the striated muscle of the glottic area prior to intubation, relaxation may be obtained by cautious intravenous injection of a rapidly metabolized muscle relaxant.

The presence of an endotracheal tube is not an absolute guarantee of an open airway. The tube may become kinked due to sharp flexion of the head on the thorax, compressed due to nasal septal deviation or obstructed due to foreign material. During any change in the position of the infant's head the tip of the tube needs only to move a short distance to be displaced upward and out of the trachea or downward into a main bronchus. Once the tube is properly placed it should be firmly anchored.

Management of the Patient During Anesthesia

Anesthesia may be maintained with a nitrous oxide-oxygen mixture supplemented with diethyl ether or cyclopropane. Light planes of anesthesia should be the rule. It is rarely necessary to use deeper than plane one of surgical anesthesia. If the use of nonflammable agents is demanded nitrous oxide may be supplemented with trichlorethylene or chloroform. Intravenous agents are not recommended unless one wishes to take into account the effects which may be exerted by them long after the operation is completed. In children under one year of age techniques should be used which do not allow the rebreathing of exhaled gases. Rebreathing is prevented by the use of valves such as the Stephen-Slater type. If partial rebreathing is allowed, as in the Gwathmey, semi-closed technique, the combined flow of gases should exceed ten liters per minute to prevent excess accumulation of carbon dioxide according to Leigh. Anesthesia for children over one year of age may be easily maintained by connecting the endotracheal tube to an ether can prepared for the Flagg technique. With this apparatus oxygen should be added to the ether-air mixture to assure

an adequate oxygen supply and to control the concentration of ether in the inspired atmosphere.

Increased intracerebral vascular tension, itself a hazard to the patient, may also complicate the operative procedure by: (1) causing the brain to swell abnormally through the open dura, and (2) by causing increased bleeding in the surgical field. The following factors, which are amenable to some degree to the control of the anesthesiologist, tend to increase intracranial pressure: (1) hypoxia, (2) hypercarbia, (3) resistance in the airway, (4) positive pressure respiration, (5) reflex coughing, and (6) impedance to venous return from the head.

Respiratory obstruction increases venous pressure, especially about the head and neck, and produces almost uncontrollable oozing from the surface of the wound. The increased bleeding tendency directly attributable to anesthetic drugs, *per se*, is minor in comparison to the results of the accumulation of carbon dioxide. This drug has two undesirable effects. Its peripheral action causes dilatation of the capillaries which contributes to general surface oozing. Its central effect causes vasoconstriction with a resulting arterial hypertension which further increases blood loss. Heavy premedication, deep anesthesia, curariform drugs, and obstruction of the airway act to increase carbon dioxide tension by reducing tidal volume.

Maintenance of Water, Electrolyte and Blood Balance

Water and electrolytes are often withheld prior to operation. Further loss during the surgical procedure may lead to serious depletion. The need for electrolyte replacement should be anticipated and fluids should be started before the operation is begun. The electrolyte problem is fully discussed in the chapter on pre- and postoperative care.

Blood loss during neurosurgical procedures is often difficult to estimate accurately. When it is realized that two saturated four by eight inch sponges can hold 30 cc. of blood, and that this amount represents one-tenth of the blood volume of a newborn infant, the need for replacing even this amount becomes apparent. An intravenous catheter or needle should always be properly placed before the operation begins. Blood replacement should parallel blood loss and a deficit never allowed to develop.

Controlled Hypotension

Bleeding into the operative field is probably the most pernicious of the many problems besetting the neurosurgeons. Contributions which the anesthesiologist may make toward reducing this hazard by *maintenance of normal physiological processes* have been discussed.

Techniques which drastically disturb the patient's normal attempts at homeostasis in order to make possible operations which otherwise could

not be considered have been developed. One of these is the deliberate production of hypotension to reduce bleeding. The blood pressure may be lowered by one of several methods. First, deep general anesthesia with agents such as chloroform and ether may be maintained during the need for hypotension. Production of profound depression of both central and peripheral control mechanisms attend the use of deep anesthesia, and these effects along with those affecting myocardial function must be considered when this method of reducing blood pressure is chosen. However, inhalation drugs are rapidly eliminated when the need for reversal of hypotension occurs. When normal tensions of oxygen and carbon dioxide in the blood are maintained, normal function of the heart, liver and kidneys are resumed shortly after elimination of the agent.

Second, arteriotomy with removal into a reservoir of enough blood to reduce blood pressure to desired levels has been described by Gardner. Since this is a state of hemorrhagic shock during which vital organs may permanently suffer from the consequent reduction in blood flow due to intense vasoconstriction, it seems to go beyond reasonable physiological grounds. When this technique is used, the blood pressure is restored by retransfusion into the arterial cannula from the reservoir.

A third method is the blockade of preganglionic sympathetic outflow by total spinal block as described by Griffiths and Gillies. Peripheral resistance is abolished and the associated vasodilatation causes pooling of blood distant to the operative site. Often this technique is not precise enough to provide sympathetic block without concommitant paralysis of respiration. Therefore, these patients must be cared for with intubation of the trachea and positive pressure respiration. Blood pressure is restored after the drug is detoxified or by the use of a vasopressor substance.

A fourth method to produce hypotension is by the intravenous injection of a ganglioplegic drug. Again, vasodilatation occurs, and if the patient is properly positioned, bleeding from the wound is diminished. Pentamethonium, hexamethonium and Arfonad, among others, have been used to this end. Of these, Arfonad seems to be the most dependable and the most readily reversible. It is given in a 0.1% solution by intravenous drip until the rate is found which produces the desired blood pressure. When it is time to restore normal blood pressure, the drip is discontinued, and if necessary, a vasopressor is injected.

Vasodilatation is a result of ganglionic blockade and total spinal block; hence, blood flow to vital structures is more nearly maintained. Although hypotension clearly would result in lessened arterial bleeding, capillary ooze and venous bleeding may not be affected, unless the patient were positioned with the surgical area above the rest of the body. However, extreme caution must be exercised in using the head-up tilt position as dangerous ischemia to the brain may occur.

Tachyphylaxis is a characteristic of ganglion blocking drugs, and in some patients, particularly the young age group, blood pressure cannot be reduced to the desired levels. It is not possible to predict which patient will respond to these drugs.

The application of deliberate hypotension in children by ganglionic blockade may lead to disappointment. Vasodilatation may occur, but often the blood pressure remains near normal. This would result in increased bleeding as shown by Enderby. A well administered ether anesthetic to a depth necessary to achieve a blood pressure of 70-80 mm. Hg., when indicated, we believe to be the safest method in children.

Indications

Neurosurgical indications for controlled hypotension in children are few. It may be necessary to help control blood loss and to shorten operating time in the case of highly vascular brain tumors or intracranial vascular malformations. It may be used in ligation of intracranial aneurysm to facilitate accurate visualization of the aneurysm so that clips may be properly placed.

Contraindications

Unless there is a clear, valid reason for the technique it should not be used. Furthermore, regardless of the method, the blood pressure should be lowered only for the portion of the operation where a bloodless field is required as advocated by Hale. It is not a method for the occasional operator or anesthetist. It should not be used unless blood is available for drop by drop replacement. Malignant hypertension, severe renal or hepatic disease have been cited by Little as absolute contraindications.

Complications

The complication rate following hypotension techniques is high. Reactionary hemorrhage has been most frequently reported. Slow recovery, difficulty in restoring blood pressure postoperatively, blurred vision, anuria and oligurea, thrombotic episodes and cardiac arrest have been attributed to deliberate hypotension. The complication rate rises markedly when systolic pressure is maintained below 80 mm. Hg.

Other Complications

Air Embolism

Air may enter the circulation during neurosurgical procedures in quantities large enough to cause serious distress and even death when the procedure, either diagnostic or therapeutic, is performed in the sitting or head up positions The large veins leading to the venous sinuses may be sub-

atmospheric. If these veins are opened air may rapidly enter into the circulation in lethal quantities.

The practice of using air pressure to speed up blood transfusions is extremely hazardous because of the danger of air embolism. The quantity of air injected in this manner may be so large as to be rapidly fatal.

Air embolism should be considered when there is any sudden fall in blood pressure or change in the character of respiration not readily explained by other factors. A brief period of cyanosis may be followed by the pale, waxy appearance of circulatory failure. Auscultation over the right heart may reveal a mill-wheel murmur. In this event the child should be immediately placed in the lateral position with the left side down to minimize an air block of the pulmonary and coronary vessels. Thoracotomy and manual cardiac resuscitation may be necessary.

Apnea

During surgical procedures on or near the brain stem irregular respiration or apnea may occur quickly. The neurosurgeon should be informed at the slightest change in respiration on these occasions. Positive pressure respiration with elimination of the anesthetic mixture should be begun promptly and the operation discontinued until spontaneous respiration is resumed.

The Postoperative Period

It is desirable that the pediatric neurosurgical patient be freed of the depressant effects of anesthetic drugs soon after the surgical procedure is complete. If this is accomplished, protective laryngeal reflexes, vasomotor stability and adequate respiratory exchange will act to avoid postoperative complications frequently seen in patients subjected to prolonged drug depression.

In the postoperative period laryngeal and carinal reflexes are often obtunded because of the brain lesion or intracranial edema secondary to the surgical procedure. If mucus and vomitus are allowed to drain into the lungs, dependent areas will develop atelectasis and pneumonia. Hypoxia thus produced will increase capillary permeability which is especially damaging in traumatized brain tissue. The rise in carbon dioxide tension will further augment the intracranial edema through its effects on systemic blood pressure. Edema of the brain will be further increased if the so-called "shock" or head-down position is used. Chest complications can be avoided if the patients are turned at frequent intervals, if secretions are not allowed to drain into the chest and if the airway is never allowed to become obstructed. If there is a probability that the patient will be unable to maintain an unobstructed airway when the endotracheal catheter is removed, a tracheotomy should be done in the operating room before extubation.

Blood pressure should stabilize rapidly in the post-anesthesia period. Hypertension and hypotension are both dangerous. If either condition develops the explanation should be ascertained and corrected immediately. Vasopressor drugs should be used with caution and never employed as a substitute for blood.

Hypothermia

The therapeutic use of cold in the physiological management of children for neurosurgical procedures has considerable value if the child or infant is febrile or if the circulation to the central nervous system is to be temporarily interrupted during surgery. Another use for this technique is in the recovery room management of patients who are hyperthermic secondary to head injuries or following intracranial procedures.

The extremely poor surgical risk associated with procedures upon children who are hyperthermic and cachectic has been recognized for many years. The change in metabolism and oxygen requirements follows van't Hoff's law and increases 13% for each degree centigrade rise in body temperature. Febrile children should have adequate fluid therapy and the temperature lowered at least to 38° C before anesthesia and surgical intervention are attempted.

The term "hypothermia" is defined by Wiggers as any body temperature below 36° C. One may designate as moderate hypothermia temperatures in the range of 36 to 32° C and as deep hypothermia temperatures below 32° C. When hypothermia is utilized during intracranial procedures we endeavor to maintain the temperature between 28 and 32° C. The physiological responses of the patient which occur when the body temperature falls below 26° C are unpredictable according to Rosomoff and should be avoided.

Subnormal temperature with the accompanying lowered metabolic requirements affords protection to the brain threatened by ischemia. Longheed and Kahn noted that at 30° C the cerebral metabolic rate is reduced to 50% of control values and at 25° C the rate is further reduced to between 23 and 35% of the controls.

Two factors are the basis of controlled hypothermia techniques: (1) depression of the temperature control center in the brain stem, and (2) the application of cold to the body. Depression of the temperature control mechanism is afforded by most of the general anesthetic drugs or may be produced through the utilization of phenothiazine derivatives such as phenergan or chlorpromazine. Body cooling is accomplished by placing patients in cold rooms, in freezers, in circulating blankets or in ice-water baths. When these two factors are combined the temperature can be reduced relatively rapidly. Our present practice is to anesthetize the child

and place him between the "circulating" blankets of a Therm-O-Rite unit. Cooling is begun coincident with the beginning of the surgical preparation. Temperature changes should be continually monitored by either a rectal or esophageal thermometer, the latter being preferred. Once active cooling is stopped, a continued decline of body temperature may occur. This appears more frequently in infants and children. The predictability of the extent of this drift is not certain.

Various methods for rewarming are utilized. Diathermy may be employed to warm the heart first before warming superficial structures. Some physicians prefer to place the child in a tub of water at 39 to 43° C while others return their patients to the wards or recovery room to allow them to gradually rewarm at room temperatures. With the "circulating" mattress it is possible to start rewarming the patient as soon as the surgical need for hypothermia is over. In this way the temperature may be gradually elevated several degrees by the time the surgical procedure is completed.

Shivering is one of the physiological variables which can occur in the management of a patient during hypothermia. If shivering is allowed to continue, it will compound many of the problems inherent to the technique in that it causes an increase in oxygen consumption, a build-up of acid tissue metabolites, and a generalized vasoconstriction which slows the rate of cooling and necessitates the additional use of anesthetic agents.

As cooling progresses respiratory minute volume diminishes. The magnitude of this decrease and the temperature at which apnea occurs depend greatly upon the depth of anesthesia, the anesthetic and premedicant drugs, and individual variations, precluding any valid prediction of these quantitative ventilatory responses to hypothermia. Arterial blood pH decreases as the carbon dioxide content is elevated. There is a greater solubility of gases at the lower temperatures and the carbon dioxide tension rises but little. Hyperventilation is commonly employed to prevent the adverse effects of respiratory depression.

The pulse rate declines linearly with the fall in temperature and the arterial pressure falls gradually. The bradycardia at low temperatures is apparently not vagal in origin and is not altered by atropine or vagotomy. Cardiac output decreases as the heart rate slows but stroke volume is almost normal down to 25° C. During the cooling process the electrocardiographic tracings show progressive changes. There is a prolongation of R-R, P-R, and S-T intervals. Berne has shown that the myocardium remains fully oxygenated, arterial and venous oxygen differences are normal or decreased and no oxygen debt is incurred during deep hypothermia. The majority of reports implicate depressed spontaneous ventilation as disposing to ventricular fibrillation. Assisted respiration or hyperventilation are recommended as routine procedures during deep hypothermia.

ACTH, 17-hydroxycorticosteroids and corticoids are all greatly depressed at 25-28° C. Epinephrine output is decreased tenfold at 26° C. Some of the immediate post-operative circulatory difficulties in patients following hypothermia may be related to this depression of adrenal response.

Although much of the interest in hypothermia is in protecting the central nervous system from ischemic damage the exact parameters of safe time or of damage at various intervals of time have yet to be defined.

The cerebrospinal fluid pressure and venous pressure decrease about 5.5% per degree Centigrade fall. At 25° C there is a decrease in brain volume of 4.1% resulting in an increase of 31.8% in the intracranial space not occupied by the brain.

The use of hypothermia always imposes possible complications and should only be employed when specifically indicated. The heart is irritable while cold and is especially irritable during the warming phase. Ventricular fibrillation may occur especially with deep hypothermia or hypoventilation. Peripheral nerve palsies may result from prolonged immersion in ice water. Skin and subcutaneous tissues may also show damage in the areas of maximum contact with the cooling surfaces. The rewarming period is the most critical. To rewarm the periphery will increase the tissue demands for oxygen and also increase the vascular bed while the heart is still hypothermic and therefore of low output. Circulatory failure may result.

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CHAPTER VI

Correlative Plastic Surgery

STEPHEN R. LEWIS

CORRELATION of plastic and neurosurgery is important in the managing of many types of problems that occur in children. The utilization of teamwork surgery will frequently enhance the care of the patient and speed up the final recovery in various congenital and traumatic problems. We must always remain aware of the multiple injury patient and insure ourselves that the general condition of the patient remains as the main consideration. Steps should be taken to move along the surgical route best needed for the over-all care. The general utilization of fine, meticulous, careful techniques are important for better wound closures and better wound healing minimizing later reconstructive problems.

The problems associated with body defects whether congenital or acquired have been emphasized by surgeons for hundreds of years. These defects have created problems dependent upon the area of the body involved. Much clinical and basic research has been done related to wounds, wound healing, and the management of the structural and cosmetic associated problems. Children, in general, have tissues that need particular care in handling, keeping in mind at all times the general condition of the patient, as well as the growth patterns related to function and appearance. The effects of trauma correlate well between the adult and child in many aspects inasmuch as the general anatomical and physiological manifestations are similar. However, there are differences in that the general physiology and application of techniques effect the management of traumatic defects in the child.

Anatomical Considerations

Before attempting surgery on the scalp, one should be aware of the anatomical peculiarities to that area. The blood supply of the scalp both arterial and venous run adjacent to one another except for the posterior auricular artery and vein which may be well separated from each other. The anterior part of the scalp plus a portion of the temporal region primarily receives its main blood supply from the frontal artery, the supraorbital artery, and the frontal branch of the superficial temporal artery. The

parietal branch of the superficial temporal artery supplies the middle portion of the scalp, and a combination of the posterior auricular and occipital arteries nourish the posterior aspect of the scalp. There is extensive anastomosing between the vessels up to the midline and minimal anastomosis across the midline with the opposite vascular supply. The venous return from the same areas is similar with the exception of the connecting branches of the emissary veins which pass through the skull to large venous intracranial sinuses. The immediate outer table of the skull is well supplied by a combination of vascular channels primarily known as the diploic

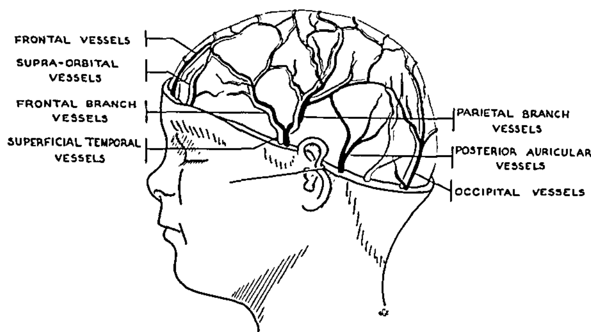


FIGURE 1. Semi-diagrammatic representation of the vascular supply of the scalp.

vessels which are between the inner and outer tables of the skull and by pericranial vessels along the periosteum. A basic understanding of the vascular pattern should be constantly a portion of the awareness of the surgeon in his responsibility to rotation of flaps or in making incisions that may markedly diminish blood supply to a part.

The skin of the scalp itself is considered as thicker than that anywhere else in the body, but again this is primarily a dermal layer, whereas the epidermis is thinner than in the general aspect of the remainder of the body. The dermal aspect of the skin is very adherent to the superficial fascia which in turn is firmly attached to the underlying occipital frontalis muscle and to the galea aponeurotica. Between these dense fibrous bands that adhere the structures together one notes lobules of fat and beneath the muscle layer the subaponeurotic layer is made up of loose areolar tissue with a loose attachment to the underlying pericranium which allows for mobility of tissue in this plane. It has been our experience that one

should keep in mind not only the type of tissue so that mobility can be directed into proper position, but also the arterial and venous supply. Adequate venous return is important to avoid congestion which in turn can cause as serious a problem of necrosis as can an inadequate arterial supply. This can be noted following dressings which are tight enough to allow arterial supply but occlude the venous return. This leads to congestion and loss of flaps.

Scalp Injuries

Trauma to the scalp, underlying skull, and brain can occur by many methods and has been of historical interest for centuries. Today, trauma has increased because of the impact of sharp or blunt objects associated with automobile accidents, industrial accidents, missiles of varying types associated with both the military and civilian increase in artillery, and many other types of injuries. Burns may be of any depth and can cause varying degrees of tissue destruction including the skull. Radiation injuries have been fairly common in the past, and one must consider that a potential atomic disaster may bring on a marked increase in the numbers of these injuries. Infections of the scalp associated with or without subgaleal hematomas can result in a severe necrosis. Resultant osteomyelitis from any of the above may pose a problem for resection of the involved area and later reconstruction.

Skin Grafts

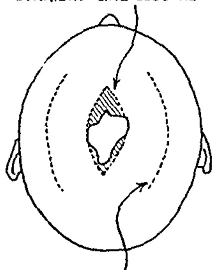
Free split-thickness skin grafts can be used on any type of defect of the scalp, skull, meninges or brain if an adequate vascular supply is present and all foreign material has been removed by adequate debridement and cleansing. Such skin grafts have the ability to give normal protective qualities of skin but usually have neither the elasticity nor the ability to withstand trauma necessary for later underlying reconstruction. Split-thickness skin grafts may be taken by calibrated mechanical instruments known as dermatomes which can be set to any desired thickness. It should be remembered that the thinner the graft, the better the take, whereas the thicker the graft, the greater the chance for loss, with other factors equal. The thinner graft will contract more over a period of time which in certain instances is desirable, leaving then the possibility of excising the graft at a later date and stretching the scalp to complete normal scalp coverage.

The success of a skin graft depends on an adequate blood supply to re-vascularize the graft, ingrowth of the capillaries, adequate hemostasis to prevent hematoma formation under the graft, and adequate immobilization to assure contact of the graft with the underlying bed at all times. Immobilization is best achieved by suturing the graft to the wound margins and utilizing a bulky pressure dressing. Usually the dressings can be

changed after the second day but it is best to wait until the fifth day if possible. Subsequent dressings are needed until total healing has occurred. Grafts take better on periosteum than when applied directly onto freshly denuded bone, although success can be obtained with direct grafts on the skull. If a take is not obtained, the underlying bone will necrose and later sequestrate. If exposed bone over a large area is present, the outer table should be removed with a chisel or perforated with multiple holes. The areas are then allowed to granulate over completely prior to skin grafting. Removal of the outer table will permit grafting at an earlier time than does the multiple hole method. Frequently skin grafts can be used in conjunction with flaps for wound closure. Grafts may be necessary to cover the bed from whence the flap came.

Use of Flaps

The next most common method for coverage is that of moving flaps either from a local area or from a distant site. The advantage of a local flap is that of providing adequate soft tissue coverage over a defect so that later bone or other material can be utilized for repair. Local scalp flaps can be undermined rapidly in the subgalea area with minimal bleeding. A flap with a single pedicle can be rotated primarily if the length of the flap does not exceed two times its width. If a longer flap is needed



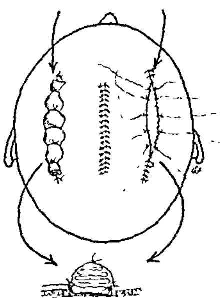
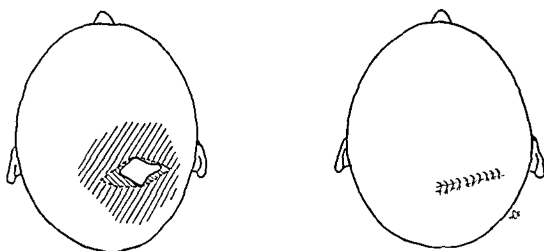


FIGURE 2. Utilization of bipedicle flaps. Either single or multiple bipedicle flaps may be used dependent upon the size of the defect

with a narrower base, it is usually preferable to make an incision and delay the blood supply along one border of the flap. This will enhance vascularization through the base and permit later transfer of the flap. A large artery within the flap allows for more security and a faster transfer time and one usually finds that if a major artery and vein are in the flap, a one-stage procedure is almost always possible. It is advisable in rotational flaps to base the flaps on the lateral aspect of the head rather than toward



DEFECT CONVERTED INTO ELLIPSE AND UNDERMINED WIDELY

FIGURE 3. The longest dimension of the defect determines the long axis of the ellipse. If the defect dimensions in all directions are approximately equal, conversion should be done vertically rather than horizontally because of the added pull of the occipito-frontalis muscle.

the midline. In handling the flaps, it should be remembered that gentleness and the avoidance of tension are necessary. The use of rotational flaps and bipedicle flaps is shown in the accompanying diagrams. Such flaps can be modified as needed in either the vertical or lateral directions. To categorically state that scalp flaps are standard in type would indicate that scalp and underlying skull defects are also standard. This is not true; therefore, one must utilize various types of flaps plus ingenuity to get adequate coverage and closure. Certain basic principles can be noted in the diagrams shown and in the various articles on scalp reconstruction that have appeared over a number of years. These may be used as reference material in planning procedures. However, in the case of immediate injury necessitating coverage, split-thickness skin grafts or local rotational flaps can be readily used for primary closure.

The third type of flap which is uncommonly used except in massive defects is the so-called distant pedicle flap which is brought to the scalp on a carrier utilizing the forearm as the carrier. The flap may either be

tubed or kept open dependent upon the type and size of defect to be covered. In children, it is better to use the tubed type pedicle flap than the open jump flap. Tubes are raised in the lines of vascularity on the abdominal wall, utilizing only the skin and underlying subcutaneous tissue. After the primary tube has been formed, a delay procedure may be necessary before transfer to the wrist as a carrier. After transfer the next procedure is a delay of the abdominal end. At this time, if desired, one can conform the delayed skin and soft tissue flap to fit the scalp defect. The next surgical procedure is to transfer the abdominal end of the flap

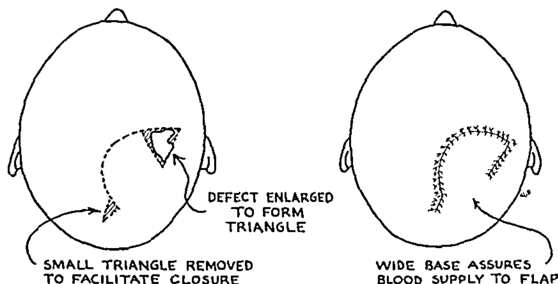


FIGURE 4. Rotational flaps showing the extension of incision from the defect in a manner to allow lengthening of a border for ultimate closure. In some instances skin grafts may be needed to cover the donor flap area to avoid tension.

directly to the scalp. The flap is maintained in place for approximately three weeks and then severed and inset. The rapidity of these flap transfers depends upon the blood supply of the flap. Associated induration or infection may delay transfer time so that no specific time interval between stages can be absolute. However, a general ruling of approximately three weeks between stages is in most instances safe. It may be found that in many instances a shorter time interval between stages is possible if all conditions are ideal.

One can note that in certain instances of avulsion of the scalp with exposure of bone if treated early enough, a rotational flap carrying blood supply to the denuded bone may protect against necrosis of the bone and later sequestration. One sees in the photographs a young child who sustained a partial-type scalp loss with an irregular defect secondary to an automobile accident. Because of the size of the defect, a large local

rotational flap was utilized for closure with very good coverage over the bone. This child has been followed for three years postoperatively and has had no further difficulty. It was necessary to use a small skin graft for the site from which the flap came. As the child grows older, this skin graft will be excised and complete scalp reapproximation obtained.



FIGURE 5. (a) Avulsed area one month after injury. A large rotated bipedicle flap with wide undermining was used for closure. (b) Postoperative result six months after surgery.

Another use for scalp flaps is in temporal alopecia or where there is loss of hair in this region from burn or radiation. In these cases a post auricular flap can be rotated forward in front of the ear to form a new sideburn. Primary closure of the donor area is usually possible with undermining. Otherwise, a skin graft can be used to cover the donor bed.

Frequently in rotating large flaps, a bulky dog-ear is formed at the ends of the flap. This should be left entirely alone for at least six weeks to avoid interfering with the blood supply to the flap. At the end of that time, by simple elliptical excision of the dog-ear, the excess tissue may be removed.

Burns and Necrosis of the Scalp

Burns may involve the full-thickness of the scalp. Similar in appearance is necrosis of the scalp resulting from extravasation of intravenous fluids,



FIGURE 6. (a) Necrotic eschar secondary to extravasation of blood. The appearance is very similar to that seen in third-degree burns. (b) Appearance two years after skin grafting. Note that the contracture of the graft has decreased the size of the original defect.

especially blood, when scalp veins are used. Photographs are shown of a two month old infant who sustained such a scalp loss secondary to an intravenous infusion which extravastated. This closely resembles a third degree burn and a very definite firm eschar is present in two areas of the scalp which comprise approximately 30% of the total hair bearing scalp region. These were excised and split thickness skin grafts used for coverage. Postoperatively she has done well and the split skin grafts have contracture diminishing the total defect. Rotational flaps and multiple excisions of the scar can be done later to give better approximation of the remaining hair bearing scalp. In burns the problem of localized infection and folliculitis may cause additional difficulty especially in the deep second degree burns where healing is extremely slow. It is recommended that shaving of the head in burns be accomplished as early as feasible. Careful cleansing and frequent utilization of saline soaks helps to keep the area clean and the granulations in a better condition for eventual skin grafting. Third degree burns should be excised and skin grafted early rather than allowing healing to occur with scar epithelization.

Congenital Defects

Congenital defects of the scalp range all of the way from mild alopecia to large complete losses with a simple membranous coverage over the underlying brain or the superior sagital sinus. The ectodermal dysplasias that are noted in the literature are surprisingly much more common in the scalp than the other parts of the body. In the small mild losses nothing need be done and waiting until the child is older is preferable before beginning reconstruction. However, where there is complete loss of the scalp and underlying skull of the membranous type, usually early drying and necrosis occurs and death ensues from either hemorrhage or intracranial infection. A case is presented of a large defect of this type which was handled in conjunction with the neurosurgeons. The child was slightly premature weighing only 4 lbs. 13 oz. at birth and on the fourth day the area was excised along with an accessory vascular sinus. Immediate coverage of the region was done with split thickness skin grafts. It was necessary to skin graft the child on another occasion over the bone where extension of necrosis occurred. Because of the lack of skull, this child is using a protective headgear until we are able to transfer soft tissue that will tolerate underlying prostheses or bone grafts. There is inadequate adjacent scalp for rotation over the entire bony defect and when the child reaches approximately the age of six or seven, an abdominal tube pedicle will be utilized for future coverage. It is interesting to note that Dr. Ira Jackson, in the workup of this child, did the routine pneumo-encephalograms which showed normal lateral ventricles with the interpretation of



Figure 7. (a) Congenital absence of the scalp twenty-four hours after birth An aberrant sinus is seen in the region of the large underlying bony defect. (b) Skin graft coverage seen two years later with the bony defect still noted in the site of cranial bulging.

a fairly substantial amount of brain tissue present. The child has progressed over a period of 4½ years quite satisfactorily although he is somewhat retarded at this age.

Scalp Tumors

Tumors of the scalp are not overly common in children. Hemangiomas rank as a frequent lesion among tumors occurring on the scalp and these are commonly of the capillary cavernous type. Many of the hemangiomas in this region are first seen in the second or third week of life and in turn gradually enlarge for a period of six to nine months when whitening may occur in the center and gradual regression over a number of years may be noted. If however, they enlarge at a rapid rate, early treatment may be necessary because of hemangio-endotheliomatous elements rather than a simple hemangioma. When surgery is needed, the majority of hemangiomas of the scalp may be handled by simple excision and closure although in some instances rotational flaps and skin grafts may be necessary.

Nevi of various sizes and shapes are also frequently observed on the scalp, forehead and face. Those that appear to have junctional activity or are cosmetically unsuitable or are frequently irritated should be excised. In the extremely large ones where primary closure is unobtainable, skin

grafts can be used for coverage. Grafts in the frontal area show increased pigmentation in a large percentage of the cases and complete forehead coverage with a thick split-thickness graft is cosmetically more desirable than a smaller graft with pigmentation changes.

Lymphangiomas, fibromas, and lipomas are occasionally found on the scalp and usually can be treated by simple excision. Eosinophilic granulomas are occasionally seen and can be handled either by surgical removal or a diagnosis made after removal of tissue for biopsy and then treatment by radiation therapy. This subject is discussed fully in Chapter XII on cranial tumors.

Radiation Problems

Radiation changes of the scalp may pose a significant problem if allowed to follow through the complete cycle of change into eventual malignancy. The most feasible therapy in our hands has been early excision in cases where radiation changes are definite and progressive and coverage with simple closure or a thick split-thickness skin graft, or if necessary by rotational flaps. It may be necessary however, in extremely extensive areas of change on the scalp, to utilize flaps brought in from a distant source. In cases where deep underlying fibrosis is present it may be mandatory to use flap coverage that carries blood supply to the part.

Scalp Incision

Incisions on the scalp for elective surgery should be carefully planned to obtain adequate exposure with minimal injury to the vascular supply and tissue. It is apparent from many diagrams in textbooks and other articles that incisions that are inadequate necessitate a new incision perpendicular to the primary one to obtain complete exposure. Where the perpendicular incision or two separate incisions meet for primary closure, frequently a central area of necrosis occurs which has to heal in by secondary intention. It is recommended that if more exposure is needed, extension of the line of incision curving it in one direction or another to maintain the maximum arterial supply is more preferable than adding a perpendicular incision.

Bone Replacement

In the closure of skull defects where there has been bone loss, the most physiological material for reconstruction is autogenous tissue. There have been excellent reports on the utilization of contoured iliac grafts or multiple split rib grafts. Tibial periosteal-osteographs have also been used for skull defects. Commonly used foreign substances are tantalum, stainless steel, acrilic and many others. Any of the foreign implants have a tendency if there is any projecting part to irritate the overlying skin and frequently

erode whereas bone has a tendency to round off and absorb in normal contour with the pressures of the skin itself. For simple filling of cosmetic defects such as in areas of the forehead, cartilage or bone has given good results and more recently the use of prosthetic materials such as polyurethane and ivalon have been successful. This subject is fully discussed in Chapter XV on trauma of the head.

Cranial Nerve Considerations

In cranial nerve or peripheral nerve problems a correlative program between specialists is frequently indicated. Those that concern the plastic surgeon most are the cases of ptosis and facial nerve paralysis. These may be either congenital or traumatic and may present a real problem from the standpoint of restoring function as well as cosmetic improvement. More than fifty different types of operations have been utilized in the correction of ptosis but basically they revolve around three distinct methods. First is that of levator shortening when there is still some function of the oculomotor nerve. Second, which includes similar basic requirements, is that of attaching the bicularis oculi to the superior rectus. The third is the utilization of nonabsorbable sutures or fascial grafts from the tarsal plate to the occipital frontalis muscle. I prefer the use of levator shortening in selected cases, and although originally described by Blaskovicks as an incision in the conjunctiva, I use an external skin incision and simply tuck the levator on itself to the desired shortened amount. In other instances, fascial grafts have been very acceptable if there is not an overly prominent supraorbital ridge. A wide strip of fascia is preferable which can be attached along the entire tarsal plate and then split into three separate strands ½ cm. above the tarsal plate so that three attachments into the frontalis muscle can be secured at a height that is desirable.

In incomplete paralysis of the seventh nerve, utilization of active exercises of the affected side in front of a mirror teaching the child to diminish the amount of activity of the normal side will often give satisfactory results. By this method, good repose and balance can be obtained and with slight smiling, a normal appearance is possible. In cases of complete facial paralysis either early repair of the facial nerve or short nerve grafts have been used. Occasionally the hypoglossal or spinal accessory nerve has been used with favorable results. The use of a portion of the masseter or temporalis muscle fastened either directly to the commissure of the mouth or to that area via fascial strips have also given good results in repose but may force a smile upon mastication. Utilization of long fascial grafts extending from the mid-upper and mid-lower lip into the temporalis fascia or muscle plus an associated tarsorraphy of the outer canthus of the upper and lower eyelids will give a good appearance in repose. In

all methods the child should be taught to minimize smiling. Newer procedures are being worked on constantly in an attempt to get more improved motion in the affected side and there is hope that the utilization of local muscles with either tendon or fascial grafts can with adequate training eventually give a better cosmetic result. One should always keep in mind in facial paralysis, the protection of the eye and primarily that of the cornea. A lateral tarsorraphy in these cases is important and should be used early before corneal damage and scarring can occur. Utilization of muscle and nerve stimulation in partial and perhaps even complete facial paralysis by competent physiotherapists should be considered as an adjunct in the therapy of not only seventh nerve injury but also in the peripheral nerve problems.

Peripheral Nerve Considerations

In the management of peripheral nerve injuries of either the upper or lower extremities, early repair is by far the most satisfactory for final regeneration. Basically, in injuries of the peripheral nerves, the type of incision or extension of the laceration for exposure is extremely important. The crossing of flexion creases in the palm, wrist, antecubital space, foot, and popliteal space should always be avoided. If the primary laceration directly crosses these creases, then the use of a z-plasty which is a local rotational flap maneuver, will allow for a zigzag closure that will break up the straight line and limit the amount of scar contracture. Incisions on the fingers for the digital nerves are best made by using the midlateral line which is the junction of the palmar and dorsal skin. One can gain adequate exposure with this type of incision and will have minimal contracture problems. In the palm of the hand, the following of the flexor crease lines may be utilized in exposing the main nerves in the hand. To cross the wrist, it is recommended that a transverse incision be made utilizing curved incisions of an "S" nature in the forearm and along the ulnar aspect of the hand to avoid flexion contractures. If in exposing nerves in the antecubital space an incision is made along the flexion crease and extending up the medial aspect of the upper arm and down the radial aspect of the lower arm, large flaps can be raised for complete exposure of all the nerves in the area. Similar type incisions are adequate in the popliteal space and leave minimal contractures. Gentle handling of the skin and soft tissues as well as the nerves themselves will produce better results than traumatic surgery. Delicate sutures with adequate postoperative immobilization will minimize the amount of scar formation and will allow for better regeneration of the nerve. In massive nerve defects of a combination of ulnar and median nerve secondary to such things as electrical burns or large avulsions, one can utilize a step transfer of the ulnar to the median nerve so as to give sensation to the major per cent of the hand.

Careful avoidance of extremes of temperature should be followed in peripheral nerve surgery until sensation has returned.

To obtain maximal function following nerve damage, early utilization of splints to maintain the hand in the proper position plus active and some passive exercises will greatly enhance the end result. Splints should be utilized until maximal fibrosis of the involved muscles of the hand and forearm have occurred. If function does not return after repairative surgery of the nerve, within a year or two, tendon transfer operations may be accomplished to restore some function. It should be remembered in the evaluation of the tendon transfers that when one group of muscles is paralyzed, the opposing group usually pulls the hand into an opposite deformity. Therefore, a balance of these muscles and their reaction is extremely necessary to get any return of function. The claw hand is seen in ulnar nerve injuries. The transfer of the extensor indicis proprius may be used to serve as the abductor when we have combined lesions of the ulnar and median nerves. The tendon "T" transplant hooked up to the wrist flexures will frequently aid in opposition of the thumb to the fingers sufficient for grasping. In cases where the ulnar nerves gives severe clawing association with loss of sensation to the little finger, amputation of the little finger will end up with a generally better hand.

We are fortunate in the number of tendons available for various transfers in peripheral nerve injuries. In cases of radial palsy the flexor carpi ulnaris and radialis can be hooked to the extensor apparatus of the fingers and thumbs by individual transfers. In median nerve injury, the transfers of the digitorum profundus supplied by the ulnar nerve may be used to activate all of the profundus tendons along with reinforcement by the supinator longus. One can add the extensor carpi radialis for the long flexor of the thumb.

Arthrodesis is a valuable procedure in the stabilization of joints and is frequently used in the proximal joints so that the distal joints may be activated by tendon motion. We have found in many children that for temporary immobilization of the position of function of the hand, which is dorsiflexion of the wrist and full flexion of the M-P joints and a 45° angle flexion of the distal interphalangeal joints, may be difficult to maintain. In these instances one can directly pin with a small Kirshner wire through the joint for temporary maintenance of position up to about three weeks without getting joint changes. However, in those cases in which we do not anticipate the eventual utilization of tendon transplants or that the tendons will not be activated and we want to get the hand in the best position, removal of the joint space and fusing the finger in a position of function will allow the child with minimal range motion to grasp and utilize the hand even with only a 10° function.

One should keep in mind the problems of Volkmann's ischemic contracture as well as local ischemic contracture in the hand secondary to injuries in the lower portion of the upper arm or in the forearm and occasionally where bandages have been kept too tight. First, and foremost, is the prevention of these two groups of contractures. Second is immediate awareness of the problem so that one can relieve the vascular insufficiency early. If, however, far advanced contracture occurs it may be necessary to slacken some of the tightened muscles, shorten the relaxed muscles, and arthrodese the wrist in a good position.

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CHAPTER VII

Neuromedical Conditions Resembling Surgical Problems

CHARLES VAN BUSKIRK

DISEASES in children which come to the attention of the neurosurgeon and do not require direct surgical therapy in general include conditions which (1) exhibit increased intracranial pressure, thus suggesting an expanding, obstructive or traumatic lesion and (2) those which exhibit a progressive history and apparent focal findings.

CONDITIONS WITH INCREASED INTRACRANIAL PRESSURE

Before discussing the causes of increased intracranial pressure which are not caused by traumatic, expanding, or obstructive lesion, a brief presentation of the factors influencing intracranial pressure seems appropriate. These factors must include the volume of the nervous tissue, itself, the volume of the cerebrospinal fluid present, the volume of the extracellular and intracellular fluid of the brain tissue and the volume of the circulating blood, both arterial and venous. An increase in amount of one or more of these factors is responsible for the increased intracranial pressure in the conditions to be discussed. The only way in which the nervous system can compensate for an increase in one or more of these various portions, since it lies in a rigid container with limited volume (the skull), is through a reduction in the amount of space occupied by the other factors listed. This, of course is not true in the very young child prior to the closure of the sutures for under these circumstances the skull itself may expand. Increased intracranial pressure thus results when the compensatory efforts of all the other intracranial elements have been exhausted. The causes of increased intracranial pressure which are to be discussed in this section include.

Cerebral edema of unknown etiology.

Dural sinus thrombosis.

Increased venous pressure due to extracranial obstruction.

Increased intracranial pressure due to pulmonary disease but not primarily due to increased venous pressure.

frequent and generally occurs in later childhood. This most commonly follows otitis but other sources of infection, including scalp or sinus infection, septicemia or meningitis, may serve. At the time of diagnosis evidence of an active infectious process may or may not be present.

Etiology

The etiology of marantic dural sinus thrombosis is secondary to the causes of thrombosis at any site, namely injury of the lining of the sinus, changes in blood which favor clotting, including dehydration, anemia, etc. and slowing of the blood stream due to cardiac factors. This type of dural sinus thrombosis is frequently associated with dysentery, diarrheas, anemias and nutritional disturbances. Chronic illnesses such as tuberculosis, malignancies and cachexias of all types may also play a part in etiology. Congenital heart disease and polycythemia have been reported to have caused spontaneous sinus thrombosis. Trauma may also be responsible. In the older age group sinus thrombosis may be coincident with an otitic infection. It may follow an otitic infection after the subsidance of inflammation and it may appear for no known reason.

Pathology

Grossly, the brain exhibits diffuse swelling accompanied by venous engorgement. Hemorrhages may be present, especially if the thrombosis in the longitudinal sinus extends into the cerebral veins. Should the clot extend to occlude the anastamosing superior cerebral veins, very extensive hemorrhages and brain softening results.

Symptoms and Signs

The symptoms which this condition may present are variable, depending upon the extent of the thrombosis. If the thrombosis is confined to the posterior part of the sinus and does not extend beyond the collateral veins directly connected to the sinus, only increased intracranial pressure with its attendant symptoms results. These include vomiting, dizziness, headache, tinnitus, and diplopia. In other cases with more extensive thrombosis, the onset may be very definite and abrupt with loss of consciousness and the appearance of paralysis and convulsions. The convulsions may be bilateral or unilateral. The signs related to increased intracranial pressure alone include sixth-nerve palsies, papilledema, and possibly optic atrophy in the long-standing case. In others with extensive thrombosis there are signs of focal cerebral deficit. Typically, there may be a cerebral paraplegia with loss of voluntary control of the bladder. The arms may be involved to some extent. Hemiplegia, however, is more common. The fontanelles may bulge and the superficial veins of the scalp may be distended. Patients

with an infectious form of sinus thrombosis may also present signs of meningitis, stiff neck, and fever.

Laboratory Findings

Invariably, there is increased intracranial pressure. The spinal fluid may reveal increased white cells, if infection is present. There may be xanthochromia or red cells present but not necessarily. The protein is generally elevated but this is not always the case. The ventriculogram may reveal ventricules which are normal, smaller than normal, or slightly enlarged. The EEG may be normal or reveal diffuse slowing.

Diagnosis

The possibility of sinus thrombosis should always be considered in infants that develop cerebral symptoms in the course of severe anemias, nutritional disorders, or other debilitating diseases. Sinus thrombosis should also be considered with the appearance of increased intracranial pressure with, or following, otitis. In the latter situation brain abscess must be ruled out. A ventriculogram is frequently necessary to arrive at the correct diagnosis. At times a sinogram is also indicated.

Course and Prognosis

Recovery from thrombosis of the posterior portion of the superior saggital sinus or from a lateral sinus thrombosis depends upon development of collateral pathways for venous drainage. Recanalization may at times occur but existing veins play the major role in providing such drainage. Anastamotic venous channels which are able to function in this fashion include anastamosing superior cerebral veins which may short-circuit about the obstruction; veins within the falx connecting the superior and inferior saggital sinuses; the scalp and deploic veins which drain into the external jugular system; veins extending forward to the orbital facial veins; and the anastamotic veins over the inferior surface of the cerebrum which connect with the transverse, cavernous and petrosal sinuses. The prognosis in patients who develop focal neurologic findings is poor, since such findings imply significant extension of the clot into the cerebral veins, thus materially limiting development of collateral venous drainage. In the presence of signs of infection, the prognosis is equally guarded. In the absence of focal neurologic findings and signs of infection, spontaneous recovery is the rule.

Extracranial Obstruction of Cerebral Venous Drainage

Elevations in venous pressure may also produce increased intracranial pressure and engorgement of the retinal veins. Papilledema, however, from this cause is extremely rare.

Etiology

The most common cause of increased intracranial pressure due to elevated venous pressure is heart failure. Another common cause is obstruction of the superior vena cava. The latter may occur as a result of phlebitis, scar tissue, tumor, aneurysms, or infections. The most probable cause of this increased intracranial pressure is an increase in the bulk of the venous blood within the head. It is doubtful if there is actual edema of the brain or an increase in the volume of spinal fluid secondary to decreased absorption. It has been suggested that since the cerebrospinal fluid pressure is elevated, along with venous pressure, these pressures balance each other and accumulation of fluid in the tissues of the brain does not occur.

Symptoms and Signs

In heart failure the symptoms presented by the patient are only those of heart failure and it has been generally noted that there is a remarkable absence of any symptoms referable to increased intracranial pressure. However, in occlusion of the superior vena cava there are, in addition to those symptoms related to elevated venous pressure of extracranial veins, such as edema, cyanosis, dyspnea and epistaxis, symptoms which are related to the nervous system. These latter symptoms are those which might result from increased intracranial pressure, such as headaches, vertigo and tinnitus. Somnolence, loss of consciousness, and seizures have also been reported. The signs present in vena cava obstruction include cyanosis of the upper chest and head and edema of the face, arms and upper chest. Other findings include markedly distended veins over the anterior portion of the chest; prominent, staring eyes, and engorgement of the veins of the conjunctiva. The fundi invariably reveal distention of the veins but papilledema is absent. Papilledema is said not to occur due to near equal pressures in the central retinal vein and in the sheath of the optic nerve.

Laboratory Data

There is greatly elevated spinal fluid pressure, frequently measuring well over 500 mm. of water on lumbar puncture. There is, in addition, an elevation of venous pressure and it has been noted that the ratio of spinal fluid to venous pressure is about 1.6. This ratio is apparently maintained both with normal and with elevated pressures. It has been observed additionally that spinal drainage will produce a fall in venous pressure. The spinal fluid exhibits normal cell count and sugar content but may reveal a markedly elevated protein. The EEG has been reported as normal.

Diagnosis

The diagnosis is not difficult, because of the concomitant symptoms and signs of heart failure or of great vein obstruction.

Course and Prognosis

The course and prognosis of this condition is related to the underlying disease producing the increase in venous pressure. The management is directed towards treatment of that disease.

Pulmonary Disease

Increased intracranial pressure and papilledema has been described in patients with *chronic pulmonary insufficiency.* This increased pressure may be secondary to some of the various processes which are associated with chronic pulmonary insufficiency, including heart failure, polycythemia, hypoxia and hypercapnia. It is believed that this syndrome of increased pressure and papilledema results from heart failure and increased venous pressure when hypoxia and hypercapnia are also present. It has been suggested that this particular combination of factors results in edema as well as congestion of the brain and that, in turn, papilledema appears.

Symptoms and Signs

Symptoms with which these patients present include dyspnea, cyanosis, as well as the nervous system symptoms of headache, impaired consciousness and tremor. The headache is frequently a prominent symptom which may be generalized or located in front or occipitally. This is persistent and of a steady, aching nature. It tends to occur most frequently at night or early in the morning. Vomiting also may occur. The drowsiness, or loss of consciousness may occur at any time, even when the patient is at work, eating, or in conversation. The patient may be forgetful, irritable, and easily confused. The signs found on examination, in addition to the papilledema, are those of tremor and twitching of the extremities. The tremor is a coarse, twitching movement resulting from an inability to maintain the extremity, the hand particularly, in a fixed posture. Electromyographically, this twitch is the result of loss of motor movement, rather than the actual contraction of muscle It is indistinguishable from that seen in hepatic coma Other signs such as dyspnea and cyanosis are present, which are secondary to either heart or pulmonary failure and are unrelated to the nervous system.

Laboratory Data

Cerebrospinal fluid pressure may be over 500 mm. of water. The spinal fluid protein may be normal, or very slightly elevated, with the normal number of cells and normal sugar content The EEG may be abnormal with diffuse slow activity seen throughout. The venous pressure is elevated and often blood ammonia is elevated. Blood gas studies reveal lowered oxygen tension and elevated carbon dioxide tension.

Diagnosis

The diagnosis is made by demonstrating significant cardio-pulmonary disease in the presence of headache, papilledema, drowsiness and confusion. This must be differentiated from other diffuse cerebral diseases, including encephalitis, lipoid storage diseases and hepatolenticular degeneration.

Course and Prognosis

The course and prognosis is dependent on the pulmonary and cardiac disease present. Treatment is directed toward correcting the underlying pathological physiology. This includes treatment of the heart failure, the use of diuretics and hyperventilation to correct the hypoxia and hypercapnia.

Lead Encephalopathy

Lead poisoning is one of the most common forms of heavy metal intoxication and is capable of producing symptoms in children which may closely simulate the clinical picture of brain tumor.

Etiology

Lead poisoning in children is most frequent in the first three years. It is most commonly the result of ingestion of substances containing lead, such as plaster and paint. Another less frequent source is inhalation of lead laden dust or fumes such as may result from burning discarded storage batteries. Severity of symptoms resulting from ingestion of lead depends to a large extent upon the amount taken and the length of time over which the lead was ingested. The most severe symptoms appear when large amounts of lead are ingested over a short period of time. Cerebral symptoms are uncommon in adults and peripheral nervous system findings are less common in children than in adults. It has been demonstrated that young animals are much more susceptible to the ingestion of lead than are older animals of the same species. There is, in addition, an individual susceptibility or tolerance to lead in the system. It has been noted that symptoms of lead intoxication may be precipitated by febrile illnesses.

Pathology

Pathologically, the brain in lead encephalopathy exhibits a marked cerebral edema and possibly some minute hemorrhages.

Signs and Symptoms

The child becomes irritable, fretful and may exhibit various gastrointestinal symptoms, such as anorexia, constipation, vomiting and abdominal

cramps. Various mental changes may appear ranging from delirium and stupor to coma and even convulsions. The child may also complain of visual disturbances. Fever may or may not be present. Examination of the fundi often reveals choked disc or papilledema. The blood pressure is generally elevated and there may be a stiff neck present, as well as focal findings, such as monoplegia or hemiplegia. These focal findings almost invariably follow a convulsion.

Laboratory Data

Laboratory data may reveal a greatly increased cerebrospinal fluid pressure, ranging from 600 to 700 mm. of water. The protein is usually elevated and there may be a variable number of monocytes in the spinal fluid, up to 100 per cmm. Spinal fluid sugar is normal. Albuminuria or glycosuria may be present. The cause of the latter is uncertain. Peripheral blood examination usually shows an anemia with basophilic stippling of the red blood cells. X-ray findings may reveal separation of the cranial sutures. There may be bands of increased density in the growing ends of the long bones and the margins of flat bones secondary to deposition of the lead. The EEG is often abnormal, showing diffuse slow-wave abnormality. There are increased levels of lead in both the blood and urine.

Diagnosis

The differential diagnosis includes intracranial mass lesions and meningoencephalitis. Diagnosis is possible because of the blood and x-ray findings characteristic of lead intoxication.

Course and Prognosis

Prognosis in the past for lead encephalopathy has not been good. A mortality of up to 65% for lead encephalopathy has been reported with marked residuals in the survivors. Neurological residuals have included optic atrophy or blindness, cerebral atrophy accompanied by convulsive disorders, and mental deficiency. More recently, the use of calcium versinate has resulted in a marked decrease in mortality and a lessening of the severity of the residuals In the presence of markedly increased intracranial pressure, frequent lumbar punctures with the withdrawal of moderate amounts of cerebrospinal fluid, one of the shunting procedures (described fully in Chapter IX, Surgery of Hydrocephalus), or a subtemporal decompression will decrease the mortality rate and also decrease the incidence of optic atrophy and blindness.

Demyelinating Diseases

Schilder's disease may occasionally produce increased intracranial pressure and papilledema. Brain tumors must be considered in such cases

presenting with unilateral findings. *Schilder's disease,* or *encephalitis periaxialis diffusa,* is a progressive organic brain disease producing widespread demyelinization in the cerebral hemispheres resulting in mental and neurologic phenomena. The cause of Schilder's disease is unknown. Theories of etiology are those which have been presented for the other demyelinating diseases.

Pathology

Grossly, the brain may be quite normal or may show a loss of substance. Occasionally, it reveals enlargement or swelling. A gross section of the brain may reveal dilation of the ventricles and possibly some cavity formation. In the older lesions the white matter may be firmer than usual. The extent of the demyelinization may be very widespread, including almost all of the white matter but these changes usually spare the arcuate fibers and the cortex. Grossly, these changes produce softened, yellow, or colorless areas. Where the condition has become chronic these areas may feel quite solid. This demyelinization is not necessarily confined to the cerebral hemispheres, but may involve the internal capsule, the cerebellum, the brain stem and the spinal cord. Microscopically, there may be only demyelinization with sparing of the axons. However, in the more severe cases, there will be destruction of the axons and cavity formation with proliferation of glia. There is also perivascular infiltration by round cells.

Incidence

Schilder's disease is rare and appears sporadically. It may be familial in occurrence with several members in the same generation having the disease. It is possible for this disease to appear at any age but most frequently its onset is before the age of ten.

Symptoms and Signs

The symptoms and signs that appear are related to the portion of the hemisphere which is involved in the process. Classically, it has been described as beginning in the occipital lobe and extending forward in the hemisphere, thus accounting for the early appearance of cortical blindness or hemianopia. There is, however, no typical area for early involvement and, in fact, any part of the brain may be involved first. Frequently an early finding will be abnormal mental behavior with slowness of speech and disturbances of memory. Hallucinations are not uncommon. Seizures, either generalized or focal, are frequent symptoms. Other common symptoms are vomiting, vertigo, and headache Disturbed vision is characteristic. This may appear as cortical blindness, hemianopia, or possibly optic atrophy Papilledema occurs in the occasional case. Focal signs are ex-

pressed as hemiplegia, cortical sensory loss, extrapyramidal deficit or cerebellar findings. Since the pyramidal system involvement is usually bilateral, there is marked spasticity, as well as pseudobulbar symptoms with dysarthria. Opisthotonus is not uncommon. The deep tendon reflexes are usually increased bilaterally with extensor toe signs. There may be extra-ocular palsies or nystagmus present.

Laboratory Data

Blood and urine are usually normal. The spinal fluid is usually normal but there may be increased pressure or increased numbers of mononuclear cells. The spinal fluid protein is normal or slightly elevated and the gold curve may be normal or show some elevation in the mid- or first zone. The EEG is variable. It may be normal, diffusely abnormal or reveal focal abnormalities depending on the parts of the cerebral hemispheres concerned in the pathologic process.

Course and Prognosis

The course of this disease is generally progressive and may result in death within one week to several weeks. Usually, however, death will result within three years after the onset of the illness. There have been reports, however, of a more chronic course occurring with the patient surviving to twenty years of age.

Diagnosis

Differential diagnosis of Schilder's disease includes consideration of a mass lesion or of multiple sclerosis. At times, ventriculography will be necessary to exclude the diagnosis of tumor.

Hypoparathyroidism

Hypoparathyroidism is a rare condition but should be included in the group of conditions under discussion since it may be accompanied by papilledema and in the past has led to operations for brain tumors.

Etiology

Spontaneous hypoparathyroidism is of unknown etiology and has been called the idiopathic form. Possible etiologies which have been suggested are infectious or traumatic. The other form of hypoparathyroidism is that secondary to operation for some other reason, in which there is removal of the parathyroid gland. This is also rare.

Pathology

Examination of the glands in the idiopathic form may show varying degrees of hypoplasia of the glands. Hemorrhages have been described, as well as replacement of the gland by fatty tissue.

Symptoms and Signs

The symptoms and signs of hypoparathyroidism are those related to a lowered blood calcium, including muscle stiffness, cramps and twitches with carpopedal spasms involving the hands and feet. Laryngeal spasm or stridor may occur. There may be sensations of tingling throughout the body, as well as convulsions, either focal or generalized. These symptoms are made worse with excitement. Increased intracranial pressure exists and papilledema may be present. All stages of papilledema, from early choke to multiple hemorrhages, may be present. Additionally, there may be other signs of increased intracranial pressure with vertigo, vomiting, and tinnitus. In the very young there may be spreading of the cranial sutures, as well. Lenticular or corneal opacities are often present and there is abnormal development of the teeth. Candida infections of the mouth, tongue and nails are not uncommon in this condition.

Laboratory Data

There are low blood levels of both total and ionizable calcium. Serum phosphorous is elevated. There is decreased urinary excretion of both calcium and phosphorous X-rays of the skull may reveal symmetrical calcifications of the basal ganglia and may also show spreading of the sutures. X-rays of the long bones may be normal or demonstrate some increase in density. Lumbar puncture may reveal an elevated pressure of up to 250 mm. of water. Spinal fluid protein and cells are normal. The electroencephalogram is normal

Diagnosis

Diagnosis is not difficult, providing this entity is considered The presence of focal or generalized seizures with papilledema and increased intracranial pressure may easily lead to consideration of brain tumor or mass lesion as the cause of disability. Diagnosis of hypoparathyroidism is confirmed by the presence of a low calcium and elevated phosphorous in the blood.

Prognosis

The prognosis is good with treatment. The proper treatment of hypoparathyroidism will lead to the subsidance of the increased intracranial pressure and of the papilledema There may be some slight residual of optic atrophy if the papilledema is prolonged

Hypertension

Another cause of increased intracranial pressure in children is *hypertensive encephalopathy*. The etiology of this condition most frequently

is kidney damage which may be infectious, toxic or obstructive; or essential hypertension. Most commonly, the hypertension of renal origin is due to acute glomerulonephritis.

Symptoms and Signs

This condition is characterized by signs referable to increased intracranial pressure, with headache, vomiting, disturbed vision and bradycardia. The respiratory rate may be reduced and focal or generalized seizures are not uncommon. Following the seizures, weakness of a post-ictal nature may be present. Papilledema is usually present with hemorrhages and patches of exudates. Vision may be reduced markedly and even be absent. Blood pressure is elevated usually over 150 mm. of mercury. There may be coexistent symptoms related to the kidney damage, including tetany, muscular twitches, and seizures, all of which are due to a disturbance of the calcium and phosphorous ratio.

Pathology

There has been observed a considerable degree of cerebral edema, flattened convolutions and often herniation of the brain stem through the foramen magnum. These abnormalities are said to be the result of vasomotor disturbances. The transient neurological findings, including seizures, are said to be the result of vascular spasm with transient ischemia of the cortex. Although this concept is not universally accepted, experiment has shown that cortical ischemia can give rise to both generalized convulsions and localized disturbances of function.

Laboratory Data

The spinal fluid reveals increased intracranial pressure with an increase in the protein content which may be as high as 200 mg. %.

Diagnosis

In the differential diagnosis brain tumor must be included where increased intracranial pressure, papilledema, or focal seizures with residual weakness is present. Other conditions that must be considered include lead poisoning, which invariably produces hypertension. Lead poisoning may also produce kidney disease resulting in hypertension with the cerebral edema on that basis. Diagnosis can usually be made by the presence of kidney damage and increased blood pressure.

CONDITIONS WITH APPARENT FOCAL AND PROGRESSIVE LESIONS

The second section of this discussion deals with those diseases which are non-surgical which may present as focal neurologic conditions. A complete list of neurologic conditions of this type would include almost

all diseases of the nervous system. Most of these conditions, if present for any length of time, may readily be distinguished from surgical lesions. There are, however, certain conditions appearing in children whose first symptomatology may mimic a surgical lesion. A few of these are presented here.

Tuberous Sclerosis

Tuberous sclerosis (Bourneville's disease, epiloia) is a condition characterized by facial skin lesions, convulsive seizures, and mental retardation.

Pathology

Widespread changes are present, not only in the central nervous system, the skin and the retina, but the kidney and other viscera as well. These are disturbances of development and are most likely the result of a defect in the germ-plasm. Gross examination of the brain reveals that it is of normal size with nodules presenting on the surface. These nodules are firm in consistency, vary in size and are white in color. These hard nodules may be distributed deep in the brain involving both gray and white matter. They are not uncommonly adjacent to the ventricles, projecting into the ventricles as tumors. Histologically, the nodules are made up of neuroglial cells with giant cells. Histological examination of the rest of the brain reveals the presence of disturbed cortical architecture, vascular hyperplasia and heteratopias. Large glial tumors may also be present. The skin lesions have been termed adenoma sebaceum but are not true adenomas of the sebaceous glands, but rather are lesions arising from nervous tissue in the skin with an increase in the amount of connective tissue and blood vessels. The retina may exhibit small congenital tumors which have been termed phakomas and which consist of glial tissue, ganglion cells or fibroblasts. Visceral lesions include rhabdomyomata of the heart, tumors of the kidney, and cysts of the liver, pancreas, thyroid or ovaries. Other congenital malformations may also co-exist.

Incidence

This is a rare condition, having been reported to constitute 66% of admissions to a mental institution. It may be familial or sporadic in its appearance It is more frequent in males than females. The skin lesions may be present at birth but appear more commonly later in life, from the third to the sixth year. These children exhibit slow motor and mental development and seizures occur within the first two years of life, usually, but may have a later onset.

Symptoms and Signs

Presenting symptoms and signs may vary to a great extent. Focal or generalized seizures appear and they are usually present before the fifth

year of life. A small per cent of focal neurologic findings may also be present. If a large tumor is present, rather than the small nodules, there may be signs of an intracranial tumor with choked disc, as well as focal findings. The degree of mental retardation may vary to some extent, being extremely mild, absent, or very severe. Psychotic episodes may also occur. There appears to be no parallel between the severity of the seizures and the severity of the mental deficit. The skin lesion on the face may vary in its appearance from a yellow or pink raised lesion to a brownish red lesion. The size may vary up to 5 mm. in diameter. These lesions are most generally distributed on the side of the nose and over the nasolabial fold of the cheek in a butterfly pattern, being present at times over the forehead and chin. Other skin lesions have been noted elsewhere, including polyps and areas of fibrous hyperplasia which have been termed shagreen patches. Warts, café-au-lait spots and anemic patches of skin have also been described in these patients.

Laboratory Data

Laboratory data depends to some extent upon the degree of visceral involvement. For example, in the presence of renal tumors, urinary abnormalities may exist. Spinal fluid is normal, excepting in those patients who have large tumors, in which case the pressure may be elevated and the protein elevated as well Skull x-ray may reveal scattered intracranial calcifications. The EEG is usually abnormal, showing several foci of abnormality as well as seizure discharges.

Diagnosis

The diagnosis presents little difficulty, if the complete syndrome is present with tumors of the retina, calcifications of the skull on X-ray, and typical pneumoencephalographic findings of small protuberances into the ventricles

Course

The general course is progressive, with increasing seizures and increasing mental deficit. There are some patients in whom the disease does not progress and remains arrested and in these a normal life span may ensue. In the usual progressive form death occurs frequently before the thirtieth year of age, from repeated seizures or chronic infections. At times these nodules may be so located intracranially that they are blocking the normal flow of cerebrospinal fluid causing hydrocephalus and increased intracranial pressure. If so, they should be treated by one of the methods for surgically reducing intracranial pressure or as Walter Dandy had done in actually removing such a mass intracranially with reported success though not altering the ultimate course of the disease.

Syringomyelia

Syringomyelia may at times closely mimic a spinal cord tumor. It is characterized by gliosis and cavity formation in the spinal cord and presents with segmental involvement of anterior horn cells and various sensory defects. The etiology of this condition is unknown but is probably on a developmental basis, since other anomalies may also be present. It is not a hereditary condition but several cases have been reported in the same family.

Pathology

Cavitation and gliosis is seen in the spinal cord. This involvement may extend from a few segments in the cord or in the medulla to involve the entire cord. It is most commonly located in the cervical or lumbar region and its extent is variable. On appearance, the cord may be atrophic or normal in size. The cavitation destroys not only sensory, motor and autonomic cells but fiber pathways as well. It commonly involves the commissural fibers in the spinal cord. In the medulla the cavity or syrinx usually lies in the lateral reticular formation and may produce defects in the function of the fifth cranial nerve, the medial longitudinal fasciculus, the descending sympathetic fibers, as well as the nuclei of the tenth, eleventh and twelfth cranial nerves.

Symptoms and Signs

The symptoms are variable, depending upon the extent of the lesion The most common location is in the cervical region. A syrinx in this location results in atrophy of the small muscles of the hands and painless burns of the upper extremity as the usual presenting complaints Weakness and atrophy of the shoulder girdle muscles may also exist and other co-existing findings may be a Horner's syndrome, nystagmus and loss of pain and temperature in the upper extremities. This latter sensory loss is in the presence of relative sparing of light touch The loss of autonomic functions of the cord can result in trophic changes of the upper extremities, producing symptoms similar to Raynaud's phenomena. The lumbar region may be involved and this is usually in conjunction with cervical involvement but not necessarily. A lesion in this location may simulate tabes Muscles of the lower extremity and its pelvic girdle may reveal atrophy and weakness and again there is the dissociated sensory loss, of pain and temperature with relative preservation of touch in the lumbar and sacral segments Bladder paralysis may result Deep tendon reflexes may be absent in the lower extremities and the plantar response may be flexor or extensor Involvement of the medulla also is usually associated with cervical cord involvement These patients present with atrophy and fibrillation of the

tongue and loss of pain and temperature on one or both sides of the face. Nystagmus, dysphagia, and respiratory stridor may also result. Occasionally, a choked disc may be present. This is possible in the presence of some associated anomaly or tumor.

Nystagmus, when present, is generally rotary in nature. Atrophy of the muscles is a very common finding in the upper extremities and spasticity is common in the lower extremities. Sensory loss is segmental in its distribution with pain and temperature being the first lost, with preservation of light touch. These segmental zones of loss may be separated from other zones of sensory loss by normal sensory areas. Distribution of the sensory deficit is usually symmetrical but not always. Long sensory tracts may be involved. At times radicular pains may occur early in the course of the illness. Examination of the deep tendon reflexes usually reveals decrease in the reflexes of the upper extremity and elevation in the lower extremity. Trophic findings, aside from the presence of recent burns and painless ulcerations of the skin, include Charcot joints, kyphoscoliosis, club feet and rarifaction of bones are also found. The presence of symptoms similar to Raynaud's phenomena with syringomyelia has been called Morvan's disease.

Laboratory Data

The cerebrospinal fluid frequently shows a slight elevation of protein. Spinal fluid dynamics may show the presence of a block. The cell count is usually normal. The course of the condition is, as a rule, slowly progressive and may continue for many years. Death results from chronic disability, with pulmonary and urinary tract infections.

Diagnosis

The diagnosis is not difficult in the presence of dissociated sensory loss, muscle atrophy, old burns and scoliosis. Differential diagnosis must include spinal cord tumor, cervical rib, or some other anomaly such as platybasia. Platybasia and the Klippel-Feil syndrome may be associated with syringomyelia.

Friedreich's Ataxia

The other condition which may rarely be confused with a focal lesion either of the cerebellum or the spinal cord is that of Friedreich's ataxia. This is a progressive degenerative disease appearing in children involving both the spinal cord and the cerebellum.

Pathology

Grossly, the spinal cord and cerebellum may appear quite normal or reveal some atrophy. Histologically, the posterior columns reveal degenera-

tion as do the lateral cortical spinal tracts and the ventral spino-cerebellar tracts. In advanced cases there may be degeneration in the dorsal roots and peripheral nerves, as well. There is usually loss of cells in Clarke's column in the spinal cord. It has been reported that there is loss of Purkinje cells and cells of the dentate nuclei in the cerebellum. The heart may be enlarged and reveal some fatty degenerative changes with chronic inflammation of the cardiac muscle fibers.

Etiology

This is an uncommon condition which is usually familial, considered probably to possess a recessive form of inheritance. Sporadic cases, however, do occur. The onset is most frequently in the first and second decades of life and is rare in the third decade.

Signs and Symptoms

The most prominent sign is that of ataxia. The child may be slow in learning to walk or the symptoms may appear after he has walked for some time. As the condition progresses some incoordination in using the upper extremity next appears. Intention tremor may appear, as well as jerky, poorly controlled movement. Speech is often affected with disturbed articulation, either in the nature of explosive or slurred speech. Additional findings are muscle weaknesses, which in the occasional patient may progress to paralysis and atrophy. These muscle changes are most common in the distal portions of the upper extremities. There is an early loss of vibratory sense and position sense is usually impaired. Occasionally touch, pain and temperature may be reduced but this is most uncommon except in the more advanced cases. The deep tendon reflexes in the lower extremities are usually lost and the plantar response is usually extensor. At times the deep tendon reflexes will be lost in the upper extremities, as well. Nystagmus is almost always present and it may be of any type, horizontal, rotary, or oscillating. Other less common findings and symptoms are those of optic atrophy, extraocular weakness, deafness, mental retardation, and seizures.

The patient may present with heart failure and heart block due to conduction defects. There are frequently skeletal abnormalities present with pas cavus and talipes equinovarus. The great toe may exhibit flexion with a hammer toe defect. These skeletal abnormalities may be present at birth or develop later. Kyphoscoliosis is present most frequently in the upper thoracic region and it has been estimated to appear in 80% of the cases. This deformity also develops late and tends to progress.

Laboratory Data

Laboratory examinations are usually quite normal. The EKG may demonstrate a conduction defect. The spinal fluid is usually normal, both in

pressure and constituents. Occasionally, the spinal fluid protein may be elevated.

Diagnosis

The onset in childhood, the familial incidence and involvement of the spinal cord and the cerebellum with the typical skeletal deformities are the common features of this disease. The diagnosis is not difficult in the more advanced cases. There may exist not only those cases which are progressive, but abortive forms as well, in which the condition may be arrested or not fully developed and remain so throughout life.

Prognosis and Course

The clinical course is usually slowly progressive with death resulting from infections, either of the pulmonary or urinary tract secondary to chronic disability. Those patients with abortive or incomplete forms may live a normal life span.

Progressive Muscular Dystrophy

This is a primary muscular disease frequently appearing in children. It may uncommonly present with symptoms suggestive of a focal nervous system lesion.

Pathology

The central nervous system and the peripheral nerves are entirely normal. Examination of the affected muscles microscopically reveals evidence of degeneration. Early in the process of degeneration the muscle fibers may enlarge and later may split to form small fibers. There is vacuolization and hyaline degeneration of the myoplasm. The number of sarcolemmal nuclei is often increased. As the degeneration becomes more complete, muscle substance is replaced with fat and connective tissue.

Etiology

This condition is not an infrequent one and there is a strong familial and hereditary incidence of the disease. Sporadic cases are also frequent. Various genetic modes of inheritance have been recorded, both dominant, recessive, and sex linked. This most frequently occurs in childhood and usually occurs in children who have had a normal infancy.

Classification

Various classifications of muscular dystrophy have been recorded and are usually of little clinical significance. Some classification of these conditions does appear useful from the point of view of determining prognosis.

The *Duchenne type* of muscular dystrophy is characterized by an early

onset before the age of six with a rapid progression of the condition and an early death. This is the most common form and presents with enlargement of the muscles appearing before weakness is present. This type starts most frequently in the lower extremities, later spreading to the upper extremities. The face, pharynx and larynx are spared. Inheritance in this type is of a sex-linked nature.

The *fascioscapulohumeral type* is next most frequent. It is variable in its age of onset from two years of age to sixty. Abortive forms are not uncommon. There is a slow progression of the condition and a normal life span is the rule. This type frequently begins with weakness of the shoulder girdle and shows, later, extensive facial involvement. The inheritance is usually recorded as a dominant form.

The *limb girdle type* usually begins in the second or third decade of life and involves most frequently the shoulder girdle and then later the pelvic girdle muscles. It may spread to other parts, but spares the face. This condition shows a slow progression and a recessive mode of inheritance is usual.

Signs and Symptoms

The symptoms presented by muscular dystrophy are those due to muscle weakness. As a general rule the more proximal muscles are more severely involved than are the distal muscles. It is difficult for these patients to raise their arms above their heads because of the shoulder girdle weakness and because of the hip girdle weakness they characteristically exhibit a waddling gait. There is no pain present and no sensory disturbance is noted. A mask-like expression is noted in those patients with facial muscle involvement. Laughing and crying may occur with impaired expressive movements and it will be noted that the eyes are incompletely closed when asleep At times there may be involvement of the extraocular movements and muscles of mastication, as well as the muscles of the palate and the pharynx, but such involvement is uncommon. Heart failure with disease of the cardiac muscle has been reported The most common area of involvement is the trunk and the extremities. Because of weakness of trunk muscles lordosis may occur In addition to the waddling gait, a steppage gait may occur from foot drop. These patients exhibit a characteristic pattern of activity when attempting to arise from a lying position on the floor They first come to their hands and knees and then to an erect position by bracing their arms and hands against their lower extremities, in a fashion which has been described as "climbing up the lower extremities" All this is a result of weakness of the hip and trunk muscles. Enlargement or hypertrophy of muscles, when it results, may present the picture of the infant Hercules The hypertrophied

muscles feel firm and rubbery on palpation. Distribution of the weakness is usually symmetrical and sensation is normal. The deep tendon reflexes are lost and the plantar response is usually flexor. Rarely, it may be extensor. There is no evidence of fasciculations.

Laboratory Data

Routine laboratory data concerning the blood, urine, and spinal fluid is normal. It has been noted that there is an increased excretion of creatine in the urine and a decreased excretion of creatinine. This decrease in creatinine is proportional to the amount of muscle which has been lost and results from a loss of ability to store ingested creatine. It has been reported that pentoses, chiefly ribose, may be increased in the urine in these patients and also that serum transaminase levels are elevated. Response of the muscles to faradic and galvanic stimulation is reduced and there is no reaction of degeneration.

Prognosis and Course

The course of this disease is variable. In general those patients who have an onset later in life are more apt to have a slower progression or may experience an actual arrest of the condition. Contractures are frequent. Death, when it occurs, may be due to heart failure from cardiac muscle involvement or due to pulmonary or urinary tract infection resulting from the marked motor disability.

Diagnosis

The diagnosis is based upon a familial history, the presence of muscle weakness at an early age, pseudo-hypertrophy, and the characteristic distribution of the weakness.

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PART II

CHAPTER VIII

Congenital Malformations

William F. Meacham

STRUCTUAL defects of congenital origin occur with such frequency in the nervous system that they constitute a major effort in the management of clinical pediatric problems. It is estimated that approximately 60% of all congenital malformations involve the nervous system, its meningeal coverings, or the adjacent bony structures. The exact cause of incomplete or delayed development of these structures is not known. While there exist many theories and speculations concerning the occurrence of such malformations, they remain principally of academic interest insofar as the practical management of these problems is concerned.

The development of the central nervous system by invagination of the primitive neural ectodermal plate to form the neural tube occurs by the third week of gestation. Beginning in the mid-dorsal region of the 7 Somite embryo the closure of the neural tube occurs progressively in caudad and cephalad directions until complete invagination occurs in the fourth week (25 Somite stage) with closure of the posterior neuropore. The mesodermal layer at this stage has become organized to form the meninges, spine, skull and their associated connective tissues. It becomes obvious, therefore, that a multiplicity of disorders of development involving the neural axis and its integuments may occur at any point from the most cephalad portion to the caudal extremity. Most commonly such defects occur along the spinal axis, the frequency increasing in the caudad direction, constituting the condition of myelodysplasia. Where there is no attempt at closure of the neural tube, a flat, undeveloped and, at times, virtually absent spinal cord results, usually in association with marked defects in the dura, vertebra and cutaneous tissues (araphic myelodysplasia). When incomplete or delayed closure of the neural tube occurs, the condition is termed dysraphic myelodysplasia, the best examples of which are hydromyelia and diastematomyelia

During the normal course of development and maturation of the nervous system an especially important topographical rearrangement occurs in association with growth in length of the embryo. After the third month

there is a proportionately greater increase in length of the trunk than of the spinal cord. Formerly occupying almost the entire length of the spinal axis, the terminus of the cord gradually assumes a more cephalad position. Due to the brain maintaining a relatively fixed position to its surrounding structures, the migration of the spinal cord continues until full growth is reached when its terminus is normally found at the level of the first lumbar vertebra. The spinal canal below this level contains the elongated nerve roots which constitute the cauda equina. The importance of this peculiar growth phenomenon will become obvious in later discussions.

Spina Bifida Occulta

Overt spina bifida occurs about once per 1000 births, but spina bifida occulta occurs with a frequency that might approach 20% of all births. The vast majority of simple defects of spine or lamina disclosed by x-ray examination are unobtrusive and symptomless and require no clinical consideration. As the name implies, such lesions remain undetectable save by x-ray examination, since there is no herniation of neural elements or meninges present. Clues suggesting the presence of spina bifida occulta

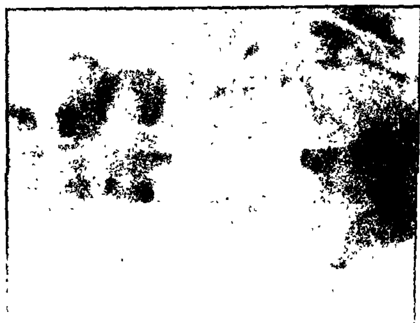


FIGURE 1. Typical radiographic appearance of lumbar spina bifida occulta

may, however, be noted on careful clinical examination by the detection of one or more of the following superficial signs: (1) localized areas of hypertrichosis near the mid-line in the lumbosacral area; (2) soft, lipomata usually irregular in size and partly moveable, but with fixation to the deeper tissues; (3) mid-line sacrococcygeal dimples or sinus tracts (which may extend into the spinal canal); (4) lumbosacral cutaneous angiomatous discoloration. Such vascular discoloration in this area suggests strongly the presence of an underlying spina bifida. Conclusive proof of the presence of spina bifida is obtained by x-ray examination of the appropriate area where defects in the lumina, widening of the canal, or fusion of the vertebrae may be detected. Minor defects in the lamina, however, may escape identification by x-ray.

The mere presence of spina bifida occulta does not constitute an indication for surgical treatment. As previously stated, the majority of such defects may remain asymptomatic throughout life and unless unmistakeable evidence of neurologic impairment exists no treatment is indicated. Since the defect occurs most frequently in the lumbosacral area, the lumbar and sacral neural elements are the most commonly involved with the resulting neurologic deficit appearing as sensory or motor deficits involving the lower extremities or as disturbances in the control and tone of the bowel and bladder sphincters. Obvious and severe paralysis of sphincters and limbs may be present at birth and cannot be considered as remediable. Milder neurologic impairment of these areas may not be suspected until such time when walking is first attempted or when toilet training is begun. Disorders of posture, faulty locomotion, disproportionate length of legs, deformities of the feet and toes, muscle atrophy, sensory loss, absence of reflexes, patulous anal sphincter, and dribbling of urine constitute evidence of possible intraspinal abnormalities requiring an appropriate investigation for spina bifida occulta.

In the event of radiographic confirmation of spina bifida associated with neurologic deficit, contrast myelography with pantopaque should be performed as a final diagnostic adjunct before consideration of surgery. The presence of an intraspinal filling defect in the immediate area of the spina bifida constitutes sufficient justification for surgery, though simple enlargement and widening of the spinal canal without obstruction or filling defect does not constitute an indication for operation and, in fact, may be construed as a contraindication, since the likelihood of improvement following surgery is extremely remote.

Surgical Treatment

As a preliminary to any operation on spina bifida occulta associated with neurologic damage the surgeon is obligated to discuss frankly and candidly

with the parents just what surgery has to offer. Since operative treatment of these lesions is primarily performed to *prevent* continued neurologic disability, parents should understand in advance that the proposed operation is not expected to restore strength and function to impaired sphincter muscles or to correct other existing deficiencies of the nervous system.

The operation is carried out in the prone position under a general anesthetic agent with tracheal intubation. The technique of laminectomy is similar to that described under the chapter on Spinal Tumors. It is advisable, however, that direct exposure of the spina bifida be deferred until an



FIGURE 2. Rachischisis. Extensive dysraphic spinal defect in cervical and thoracic areas.

area of normal spinal canal above or below the lesion has been exposed. The dissection in the region of the defect can then be approached from an area or normal structural relationships. Great care should be exercised in the removal of intraspinal lipomatous tissue which may exist both extradurally as well as intradurally. Where the meninges are deficient the identification of neural elements in this fatty tissue may be extremely difficult and if efforts to remove all such abnormal tissues are persisted in additional neurologic damage may occur. The dura should be mobilized and freed from the adjacent overlying tissues to allow for closure of the dural opening Where neural structures are adherent to the dura they should be gently dissected free. Any existing incomplete laminae are rongeured away to leave a wide bony decompression. If possible, the paraspinal muscles are re-approximated over the defect and the superficial tissues closed in layers. Stabilization of the spine by bone transplants is not indicated The operative defect resulting from laminectomy requires no special protection in either children or adults.

There is no accurate way to quantitate the results of such surgery.

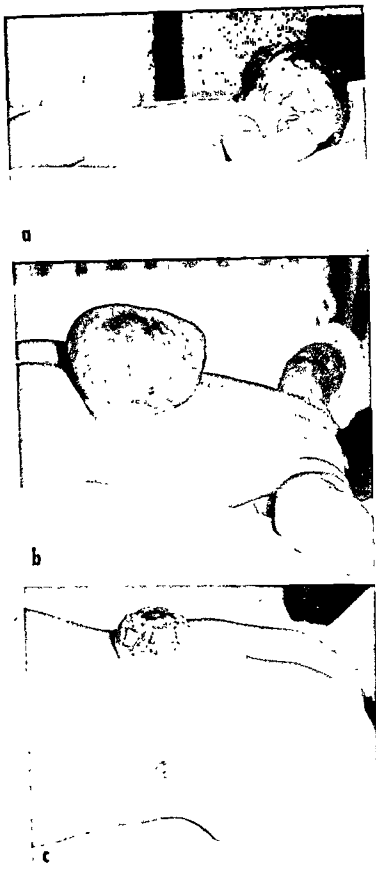


FIGURE 3 (a) Lumbosacral myelomeningocele associated with lipomatous masses about the defect. (b) Lumbar meningocele. Covering consists of thin, translucent membrane. (c) Lumbar myelomeningocele with ulceration and necrosis of the sac.

Careful evaluation of such patients over a long postoperative period may reveal improvement in an otherwise progressive disorder.

Meningocele—Myelomeningocele—Rachischisis

Since clinical nomenclature requires a differentiation in the various forms of spina bifida, these anomalies will be separately considered, although much of what has been described regarding spina bifida occulta will apply to these conditions.

In extreme cases of spina bifida the spinal canal may be wide open over a distance of many vertebral segments. The spinal cord is exposed and is subject to severe degenerative changes. Such a condition is termed *holorachischisis*, and is not compatible with life. More localized forms of *rachischisis* are synonymous with spina bifida and when associated with herniation of the meninges are commonly termed *meningocele.* When neural elements are present in the herniation the term *myelomeningocele* (*or meningomyelocele*) is appropriate. Such anomalies constitute forms of neural dysraphism.



FIGURE 4 Paralysis of anal sphincter with rectal prolapse due to myelomeningocele Note excoriation of perineal skin due to constant dribbling

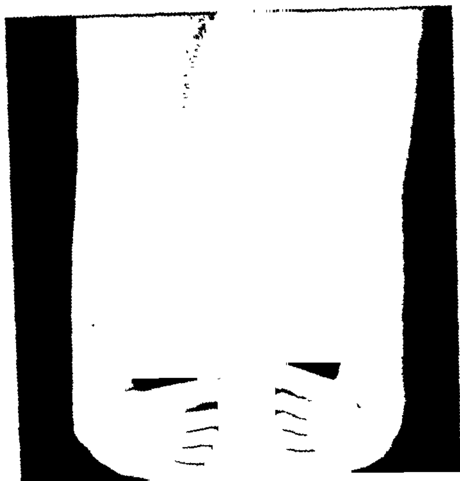


FIGURE 5. Typical deformity of lower extremities in myelomeningocele

Diagnosis

The diagnosis is obvious at birth and except for the rare sacral neoplasm which may simulate a lipomatous meningocele there is no difficulty in diagnosis. However, differentiation of meningocele from myelomeningocele may be more difficult. Each may occur at any point along the spine although the most common sites are the lumbosacral and the cervical areas. Each may be pedunculated or sessile, covered with healthy, full-thickness skin or with deficient, thin, friable epidermis. The covering may consist entirely of a translucent membrane over the dome, merging into epithelial margins at the base where the skin frequently is erythematous and pigmented. In some the sacs are composed of thick, pitted, scarred and trabeculated skin with areas of focal necrosis and ulceration. Lipomatous masses frequently cover the sac. If the lesion lends itself to transillumination, the recognition of neural elements in the sac will establish the diagnosis of myelomeningocele. If there is obvious neurologic impairment of sphincters and limbs regardless of the presence of hydrocephalus, the lesion is most certainly

effecting a more satisfactory repair due to spontaneous epithelial growth as well as a relative reduction in the size of the lesion. It should be remembered that surgery in such cases is not effective in controlling hydrocephalus nor in improving the neurologic status. On the contrary each may be aggravated by the operative procedure.

(4) Infection of the skin, meningocele sac or meninges constitutes a contraindication for surgery regardless of location, neurologic state, or hydrocephalus, until all clinical and bacteriological evidence of the infection has subsided.

(5) The development of a progressive and rapidly increasing hydrocephalus serves as a contraindication for surgical repair of a myelomeningocele regardless of location until such time as the hydrocephalus has either spontaneously arrested or has been alleviated by surgical correction of the associated Arnold-Chiari malformation. In the event of a very thin or ruptured membrane covering, however, the closure of the spinal lesion

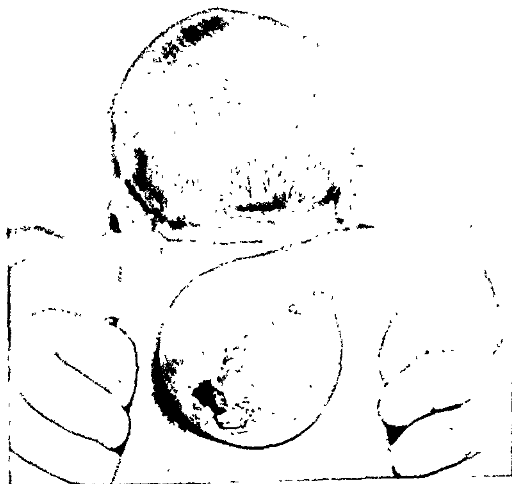


Figure 7 Pedunculated meningocele upper dorsal area. There is no associated hydrocephalus.

may necessarily take precedence in order to protect the infant from an almost certain meningeal infection.

(6) In the absence of hydrocephalus, all lesions associated with partial neurologic deficits should be considered for operation in the first three months of life if the defect is amenable to closure, otherwise further delay is advisable.

(7) Surgery is contraindicated in infants with myelomeningocele who have an associated progressive hydrocephalus with complete paralysis of bowel and bladder sphincters and lower limbs. The prognosis in such cases is virtually hopeless in terms of such a child living except as a bedridden hydrocephalic paraplegic with all the attendant miseries of sphincter incontinence, urinary tract infections, mental retardation and numerous other complications. On the other hand, if the hydrocephalus does not develop, or arrests spontaneously before becoming excessively far advanced, the paraplegic child may indicate a surprisingly bright mental capacity and become a candidate for surgical correction of the myelomeningocele at a later time for purposes of cosmetic improvement.

Occasionally early closure of leaking lesions is carried out in this clinic in such relatively hopeless cases, but only after the parents are fully informed about the true state of affairs. On their insistence, therefore, a simple cutaneous type closure of the defect is deemed advisable to prevent the demise of the infant from meningeal infection. It becomes almost a philosophical matter since it may appear that by so doing the infant may be temporarily spared, only to succumb to other complications. Were it not for the occasional instance of salvage in such cases such a clinical philosophy would be untenable.

Operation

The operation is carried out in the prone position under general endotracheal inhalation anesthesia. It is essential that a dependable intravenous cannula or catheter be in place for necessary fluid and blood replacement. The meningocele is encompassed by elliptical incisions which join above and below in the direction of the longitudinal spinal axis. In low lumbar and lumbosacral lesions it may be advisable to employ transverse incisions so that the gluteal fold area may be avoided. The skin flaps are undermined widely and mobilized sufficiently to allow for adequate exposure of the base of the sac. By sharp dissection the fatty tissues about the meningocele are divided and the dural membrane exposed near the base throughout its circumference. At this stage the size and extent of the actual defect can be ascertained. The neck of the sac may be surprisingly small in lesions that appear quite large on the surface. The isthmus is then dissected free from all attachments to bone and connective tissues. At this point the

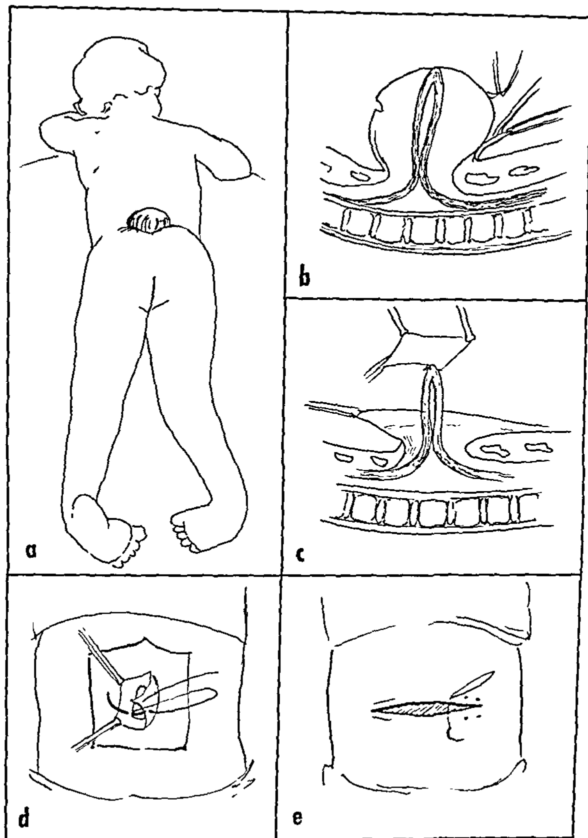


Figure 8 Stages in the repair of lumbar myelomeningocele.

sac is opened near the apex and the contents carefully inspected after excising redundant portions of the sac and surface tissues. Neural elements that occupy the sac and are adherent to its inner surface are gently freed and placed within the spinal canal. Where adhesions between the membrane and nervous tissues are too intimate to allow such separation that



FIGURE 9. X-ray appearance of extensive lumbar spina bifida with large myelomeningocele.

portion of the sac is allowed to return to the spinal canal with the nerves, since even the most careful dissection might result in increased neurologic damage.

Excessive portions of the dural sac should be excised, care being taken to preserve enough of the neck to allow for a water-tight, non-constricting closure with fine silk. At times it may appear that the neural structures have forfeited the right of domicile within the spinal canal, but with

gentle persuasion their reposition can be accomplished. The bony defect is left undisturbed and attempts to employ bone grafts are to be discouraged. Wherever possible, however, the defect should be covered by the imbrication of leaves of reflected lumbodorsal aponeurosis. Careful closure of the skin and subcutaneous tissues is most imperative. Skin closure under tension is to be condemned since it may result in loss of viability of skin margins resulting in slough and infection. Sufficient skin mobilization is ordinarily obtained by wide undermining of the flaps, but occasionally, peripheral, relaxing incisions may be necessary. The wound is covered with flat gauze and completely sealed by waterproof adhesive.

Postoperatively, the child is kept on its abdomen on a pillow with the lower limbs dependent so that urine and feces may fall on a folded diaper placed between the legs. The dressing is left undisturbed so long as it remains clean and dry, until the sutures are removed on the seventh day. Significant elevations of temperature may indicate the necessity of periodic inspection of the wound for signs of infection.

Usual nursing routines are allowed after the first postoperative day. The baby may be weighed daily, bathed, and held for its feedings. Restraints are not necessary and with so little restriction of movement there is little chance for the development of pressure areas. Growth of the head is noted by daily head measurement of the fronto-occipital circumference. This is continued by the family at weekly intervals after the child is discharged from the hospital. Associated deformities of the feet and lower limbs, so commonly accompanying myelomeningocele should be treated early and concomitantly with the spinal lesion. The correction of such deformities by enlisting orthopedic advice is advisable. The value of a "team" consisting of pediatrician, orthopedist, neurosurgeon, urologist and physical therapist should be obvious.

Encephalocele

Like the various lesions associated with spina bifida, those occurring with cranium bifidum are classified according to the contents of the sac. Where no neural tissue is present the lesion is termed a *cranial meningocele*, otherwise the term *encephalomeningocele* is proper. Common usage, however, groups all such deformities as *encephaloceles*. Occurring less frequently than spinal lesions, they constitute lesions commonly seen in large clinics. Like meningoceles they may occur in all sizes and shapes and with various types of covering, ranging from a thin membranous sac in imminent danger of rupture, to a full-thickness integument. They may be sessile or pedunculated and may occur over any portion of the skull near the midline. Encephaloceles show a predilection for occurring most frequently in the occipital area, diminishing in rate of occurrence over the parietal, frontal, nasal and orbital regions. The rare nasal encephalocele may be mistaken

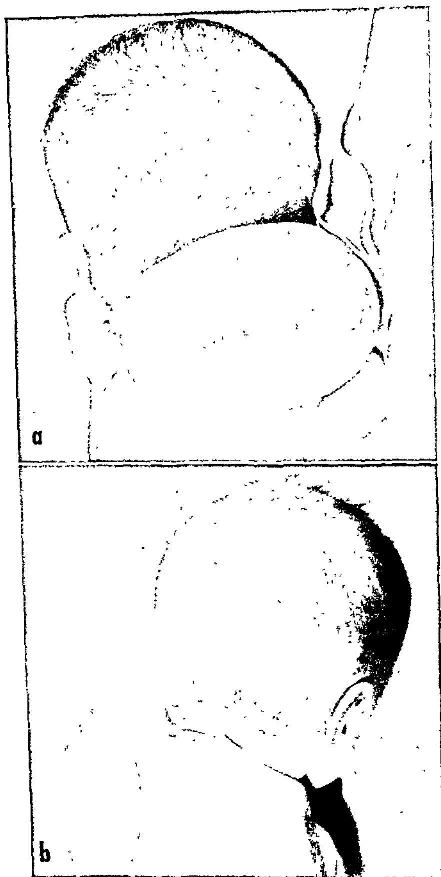


FIGURE 10. (a) Occipital encephalocele. Cerebellar tissue was present in the sac. (b) Operative wound following excision and repair of lesion in Figure a.

for a nasal polyp or other primary intranasal tumor. The disastrous effects of attempted removal trans-nasally can be imagined. Except for the orbital encephacele which may be an obscure cause of exophthalmus, the remainder are usually identified without difficulty at birth, although smaller lesions may be confused with tumors of the scalp.

By careful examination with transillumination and by palpation the presence of neural tissues within the sac can often be ascertained although the nature of such tissues cannot be determined except with operative exposure. There are no characteristic clinical signs indicative of cerebral herniation within the sac. The signs of dysraphism are frequently present as in spina bifida (hypertrichosis, cutaneous angioma). X-ray films of the skull are routinely taken in order to evaluate the size and extent of the cranial defect. In the presence of hydrocephalus or gross brain involvement

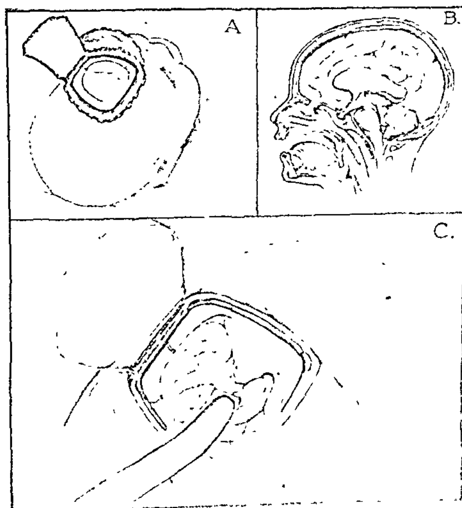


FIGURE 11. Nasal Encephalocele. (A) Frontal craniotomy approach. (B) Saggittal view of relationship of encephalocele to cranial defect. (C) Stalk of frontal lobe entering defect at the cribriform plate. (Courtesy of Dr C. A. Cobb and Williams and Wilkins Co.)



FIGURE 12. Small, sessile frontal encephalocele, often confused with sebaceous cyst of scalp.

there is some merit in the use of ventricular air studies which may reveal evidence of additional intracranial abnormality or reveal valuable information concerning the relationship of the intracranial structures to the encephalocele.

Since the physical characteristics of encephaloceles render most of them operable, it is wise to consider removal and repair of the lesion in the first four weeks of life unless other malformations and the general condition of the baby preclude consideration of surgery. Nasal, orbital, and frontal encephaloceles, if diagnosed early, may be deferred until later in childhood for surgical correction.

The bizarre and unsightly nature of encephaloceles makes surgical removal for cosmetic reasons almost mandatory. In thin-walled sacs where ulceration, rupture, or leakage is probable, surgical correction of the lesion

is, of course, indicated in order to prevent subsequent infection. General nursing care and management of the infant is materially enhanced by removal of the encephalocele.

Operation

The operation is carried out in much the same manner as that described for the Arnold-Chiari malformation. The base of the encephalocele is encompassed by elliptical incisions in either the transverse or longitudinal axis depending on which method seems to facilitate a convenient closure. After the scalp layers are somewhat undermined to expose the cranial defect, the neck of the sac is dissected free of the bony margins throughout its circumference. The sac is then opened and its contents examined. If neural tissue is not present the redundant portions of the sac are excised, leaving enough of the membrane to allow for a tight closure. If the cranial

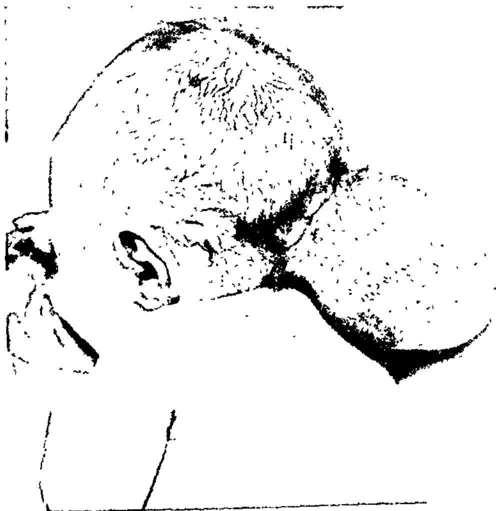


FIGURE 13 Large, pedunculated encephalocele containing cerebellar tissue at the pedicle. Large sac contains fluid only



FIGURE 14. Chronically infected and ulcerated nasal encephalocele.

defect is small a flap of periosteum can be reflected over the defect to reinforce the dural suture line.

If neural tissues are present within the sac they are freed from any attachment to the membrane and reposited within the cranium. In some instances massive tissue herniation will have resulted in obliteration of blood supply to the neural tissues with ischemic necrosis and fibrosis of large portions of the mass. Where this can be recognized as constituting functionless tissue it can be carefully excised and the remainder of the operation carried out as described above. No effort should be made to cover the cranial defect with bone, tantalum, or plastics at this time. The scalp is closed in layers over the defect.

The repair of orbital and nasal encephaloceles is carried out entirely as an intracranial procedure performed through a coronal or concealed type frontal incision. In the former instance the orbital roof is removed as in the usual orbital decompression and the dural repair effected after the

sac has been dissected free of the periorbital tissues. In nasal encephalocele herniated cerebral tissue through the cribriform plate is carefully freed and brought back into the intracranial intradural exposure where it is amputated and the dural defect closed. Surgical manipulations of the encephalocele by the trans-nasal route are to be avoided.

Intraspinal Meningocele

This form of meningocele is rarely seen in conjunction with a spina bifida and may be disclosed at operation for the latter. It consists of a thin-walled dural cystic structure within the spinal canal, separate and distinct from the dura which encloses the neural elements except for a small neck through which communication with the subarachnoid space is maintained. In older children and adults the lesion may produce extreme widening of the spinal canal with thinning and erosion of the pedicles over several segments. The anterior wall of the spinal canal may show a striking convexity on lateral x-ray view. Demonstration of the intraspinal sac may be demonstrated by myelography, particularly when the contrast medium fortuitously gains entrance to the sac. The site of most frequent occurrence is in the lower thoracic, upper lumbar areas. Disturbances of gait, and various forms of sensory and motor impairment of the lower extremities, may result from the gradual expansion of the cyst. This lesion may be synonymous with or indistinguishable from the spinal extradural cyst thought to be congenital or of traumatic origin.

Treatment consists of appropriate laminectomy with removal of the cyst and closure of the communicating neck (which may be quite attenuated).

The Arnold-Chiari Malformation

The relationship between myelomeningocele and hydrocephalus has long been recognized as a baffling and difficult clinical problem. A high percentage of children with myelomeningocele will show some degree of progressive hydrocephalus which may arrest spontaneously at any stage. Various theories relative to disturbed cerebrospinal fluid production-absorption balance have been propounded. How much the membranous meningocele sac influences the fluid absorption is not known, nor can it be quantitated. On a more mechanical basis it would seem that the anchoring effect of the displaced neural tissue within the sac may interfere with the normal ascent of the cord. This "tethering" of the cord is thought to result in a gradual traction of the hind brain caudally so that cerebellar tissue is found occupying the upper cervical canal (*Arnold's deformity*). The medulla is elongated and kinked posteriorly (*Chiari's deformity*) Whether this deformity is the result of growth traction, a primary developmental anomaly, or an overgrowth phenomenon is not known.

The severity of the hydrocephalus may be directly proportional to the

magnitude of the Arnold-Chiari deformity. It is likely that the abnormality exists to some extent in all cases of myelomeningocele. The mechanical effects of the deformity in interfering with passage of the cerebrospinal fluid from the fourth ventricle into the basilar cisternae and the spinal subarachnoid spaces is obvious.

Infants with rapidly developing hydrocephalus must be considered candidates for surgical correction of the obstructing deformity before correction of the myelomeningocele, and in those infants exhibiting an arrested or

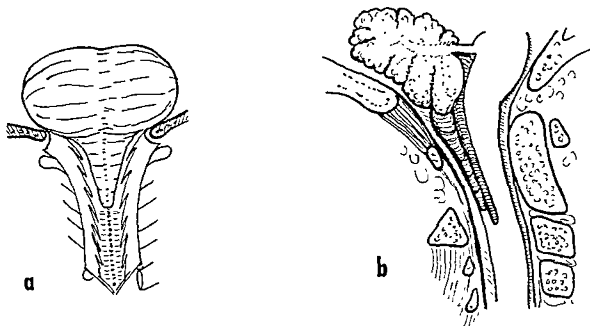


FIGURE 15. Arnold-Chiari malformation showing herniation of cerebellar tissue into spinal canal. (a) Posterior-anterior view. (b) Lateral view.

mild hydrocephalus repair of the spinal lesion is frequently followed by a pronounced activation of the hydrocephalus. The cause for this is unknown, but may be due to further impaction of the tissues in the upper cervical canal rendering the obstruction more complete. The use of constant ventricular drainage through a closed system may be helpful in alleviating the disastrous effects of the hydrocephalus until a gradual readjustment and restoration of balance occurs. At times, however, the hydrocephalus remains progressive in spite of all attempts to retard it.

It should be noted that the Arnold-Chiari malformation occurs occasionally as a solitary abnormality without evidence of any associated meningocele or myelomeningocele.

Operation

The operation is performed with the infant placed in the classical, face-down cerebellar position. General endotracheal anesthesia is used. Through a generous mid-line incision extending from the inion to the lower cervical

region, the occipital bone, foramen magnum and the upper four cervical spinous processes and laminae are exposed. A suboccipital craniectomy is performed over an area sufficient to expose the midline cerebellar structures as well as the medial portions of the cerebellar hemispheres. The lamina of the upper three cervical vertebra are removed and the dural opening is made in the lowermost portion of the spinal exposure and carried upward to the level of the foramen magnum. At this point the dura over the cerebellum is incised laterally on each side in a stellate fashion back to the margins of the bony exposure. Troublesome bleeding is usually encountered from dural vessels in the form of a widespread venous plexus in the dura overlying the cerebellum. The spinal dura is retracted widely and sutured to the lateral muscles, to be left unclosed. Tongue-like extensions of cerebellum are seen covering the lower medulla and cervical cord, usually quite adherent to the cord as well as to the adjacent dura. If the caudal extent of this tissue extends below the level of the exposure, additional lamina are removed until the distal extent of the herniated tissue has been reached. Separation of the external adhesions is done with care to avoid bleeding. The lateral posterior angles are similarly exposed on either side of the medulla. Elevation and amputation of the abnormal cerebellar tonsils may be tempting but is unwise since complete removal from the adherent spinal cord and medulla may be associated with troublesome bleeding and unjustified trauma. The distal portion of the fourth ventricle is identified by elevation of the vermis or by splitting the lower portion of this structure. If a free flow of fluid from the fourth ventricle ensues the wound is then closed in layers with non-absorbable sutures. Where free communication of fluid from the exposed fourth ventricle cannot be demonstrated it is proper to assume that a stenosis or atresia or the cerebral aqueduct exists In such a situation a diversionary type of shunting procedure is employed The usual Torkildsen operation (ventriculocisternostomy) cannot be used due to the absence of a well defined cisterna magna, and must be modified so that the distal end of the tube is placed well down in the anterior spinal canal or into the cisterna lateralis. A ventriculo-pleural, or ventriculo-peritoneal shunt may be preferred. Due to the genito-urinary complications associated with compromised bladder sphincters, the employment of a ventriculo-ureteral shunt is contraindicated.

At best, there is little room for optimism over the results in such severely afflicted children

Internal Hydrocephalus

In the clinical sense, the term internal hydrocephalus indicates the accumulation of fluid under pressure in the ventricles of the brain. A full treatment of this condition as a clinical entity will be considered under

Chapter IX on Hydrocephalus although hydrocephalus in connection with spinal bifida, myelomeningocele, encephalocele and the Arnold-Chiari malformation has already been alluded to. Since hydrocephalus is the outstanding clinical feature of two other abnormalities it is relevant that these should be considered here in some detail.

Atresia of the aqueduct of Sylvius may be present in several forms. Pathologically, the aqueduct may be *completely absent as a functional* lumen and is identifiable by the persistence of a few clumps of ependymal cells, or be a few small, minute cysts lined with ependyma. On the other hand the aqueduct may be well formed but occluded by a proliferation



FIGURE 16. (a) "Setting sun" phenomenon associated with advancing hydrocephalus. (b) Enlargement of head with distention of scalp veins due to hydrocephalus (atresia of cerebral aqueduct).

of ependyma or by gliosis. Rarely, the aqueduct may be perfectly formed *but occluded by a thin membrane or transverse band probably representing* the remnant of a localized, segmental area of atresia which has gradually become stretched by hydrostatic pressures. There is no known cause for congenital aqueductal obstruction except in some cases there may be a hereditary factor present.

The diagnosis is made during the investigation of hydrocephalus which may be noted at birth, or becomes obvious in the early months of life. The obstructive nature of the condition becomes obvious with the performance of the dye test as described by Dandy wherein dye injected into the lateral ventricle is not recovered promptly by lumbar puncture.

A more accurate demonstration of the site of the obstruction is obtained by ventricular air injection where the failure of air to pass through the aqueduct into the fourth ventricle can be recognized. If doubt exists regarding the site of the obstruction, the posterior fossa should be explored and the presence of patency of the aqueduct established either by direct inspection of its distal end or by failure of a small, soft catheter to pass freely

Treatment consists of correction of the obstructive hydrocephalus by the use of an artificial exit from the ventricular system proximal to the obstruction. This is best performed by the use of a by-pass shunt from the lateral ventricle to the cisterna magna or anterior upper cervical subarachnoid space (Torkildsen's ventriculocisternostomy). Providing the basilar cisterns are developed and functional an excellent palliative result may be obtained, otherwise additional shunting procedures may be required to arrest the hydrocephalus.

Congenital atresia of the foramina of Luschka and Magendie produces a peculiar type of obstructive hydrocephalus. Here the entire ventricular system is dilated tremendously with no functional exit for the ventricular fluid at the outlet of the fourth ventricle. As a result the posterior fossa becomes the reservoir of a tremendous cyst representing an enormously

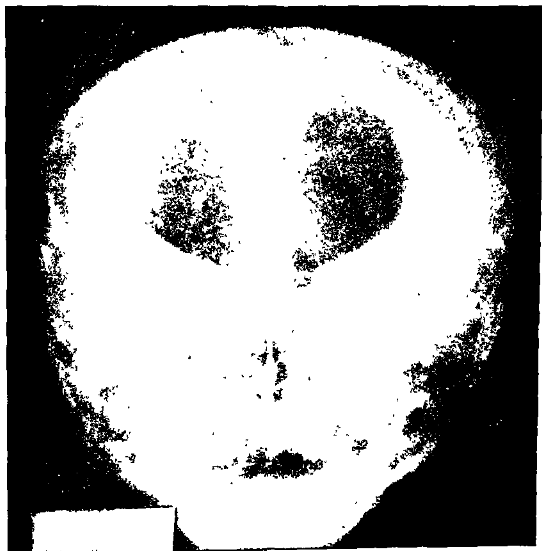


FIGURE 17. (a) Congenital atresia of foramina of Luschka and Magendie. Note large fourth ventricle with small nubbins of cerebellum Common lateral ventricles.



FIGURE 17. (b) Lateral view of (a) showing huge "cyst" occupying posterior fossa.

distended common fourth ventricle and cisterna magna. The lobes of the cerebellum remain as rather primitive nubbins of tissue and the vermis may be absent or represented only by a transverse bar or ridge which is pushed far forward. The medulla and floor of the fourth ventricle is flattened and widened so that the emerging cranial nerves seem to arise from beneath the flattened brain stem. The cerebral aqueduct is shortened and dilated to a degree that suggests a foraminal opening rather than an aqueductal passage. The tentorium is thus displaced upward by the presence of the "cyst," with the torcular and lateral sinuses similarly dislocated.

Clinically the condition is suggested in a hydrocephalic child possessing a high inion, and who may show radiographic evidence of upward displacement of the posterior dural sinuses. The Dandy test will reveal the obstructive nature of the lesion and intraventricular air studies will disclose the presence of the huge cystic lesion in the posterior fossa.

Operation

The only treatment is surgical. Through a midline exposure a suboccipital craniectomy is performed along with the removal of the arch of the atlas.

The dura is opened over the exposed area in stellate fashion to expose the membrane of the cyst which is then opened and carefully trimmed away throughout the periphery of the exposure with particular attention to removal of the membrane at each lateral angle. Communication with the basilar cisternae and spinal subarachnoid space is now theoretically possible. The dura is allowed to remain open and closure of the wound is carried out in the usual manner. Relief of intracranial hypertension with arrest of the progressive hydrocephalus will constitute an excellent result since cerebellar ataxia and disturbances of coordination may remain as a permanent neurologic deficit. If hydrocephalus persists or recurs due to failure of cerebrospinal fluid absorption additional shunting operations may be indicated.

Diplomyelia and Diastematomyelia

Diplomyelia constitutes a congenital malformation characterized by a duplication of the spinal cord for a variable number of segments. In more than one-half the cases it is associated with spinal bifida. There may be

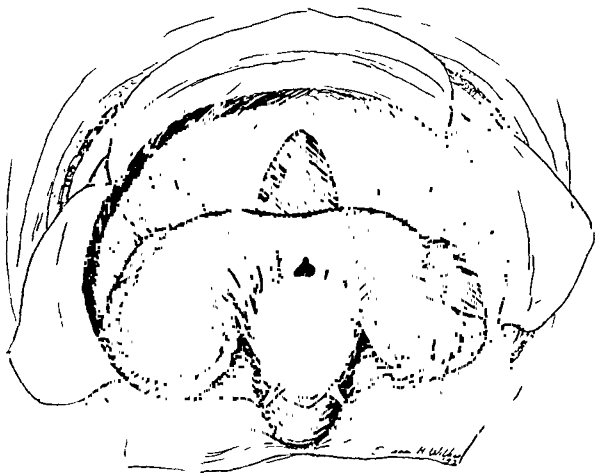


FIGURE 18 Appearance of rudimentary cerebellum and flattened medulla in congenital occlusion of foramina of Luschka and Magendie

two separate canals with a more or less normal arrangement of gray and white matter about each. A separate pial investment is present for each structure but the dura and arachnoid serve as common coverings. There are no specific clinical findings associated with diplomyelia, the condition often being discovered in operations for spina bifida or as a finding at autopsy.

Diastematomyelia represents a form of dysraphism occurring in the low dorsal or lumbar region as an occult malformation, in association with some defect of the vertebra, usually spina bifida. The condition is to be differentiated from diplomyelia since a simple splitting of the cord occurs rather than a true duplication. Presumably due to a mesenchymal cell rest a spicule of cartilage or bone is formed which acts as a transfixing septum through the cord or cauda equina. Such fixation results in a low anatomical position of the cord and impairs the normal ascent of cord, which by continued growth against the spicule may result in a cleft in the cord extending over a distance of several segments. Each portion of the divided cord is invested in its own dural compartment.

The disorder may be suspected in cases of spina bifida with the usual cutaneous abnormalities and with neurological disorders involving the sphincters and lower extremities. The diagnosis cannot be made with certainty except by x-ray confirmation of the presence of a midline bony spur. The spinal canal is frequently widened and with the use of myelography the septum can be visualized as a filling defect in the area of the canal corresponding to its widest portion. When the diagnosis is established operation should be recommended as a measure to prevent further neurologic damage.

Operation

The operation is performed as in any standard laminectomy with a full segment exposed above and below the bony spur. The dura is then gently separated from the spur and the spicule is carefully nibbled away with a small rongeur. The dura is opened by incising it above and below the split, the incisions joining at the cleft to excise the dural septa. The remaining portion of the spur can then be removed down to the anterior wall of the canal, following which the posterior dural opening is closed so that the cord is then enclosed in a single dural envelope which remains open on its anterior surface. The remainder of the wound is closed in layers with non-absorbable sutures.

Congenital Scalp Defects

Congenital defects of the scalp consist essentially of localized areas of absence of the scalp and while usually small in size the defect may be so

extensive as to pose a difficult problem in plastic repair. Such lesions may mark the site of an underlying congenital defect of the bone and when small in size no treatment other than excision and closure of the scalp lesion is necessary. Large defects of the scalp require early definitive excision and closure if at all possible. Unless the infant is treated shortly after birth the scalp defect will be converted into a mass of hard, dried secretions covering an inspissated membrane. Regardless of the presence or absence of an underlying bone defect, the lesion should be excised to prevent bleeding and infection and the defect kept clean and moistened until spontaneous growth has covered it or until a plastic correction of the defect can be accomplished. The treatment is fully discussed under Chapter VI on plastic surgery.

Congenital Skull Defects

Congenital defects of the skull (other than forms of cranium bifidum) consist of persistent parietal foramina which contain a persistent emissary vessel and congenital absence of portions of the skull. Persistent parietal foramina require no surgical consideration as far as the skull defect is concerned, although the distended, anomalous parietal emissary vein may require ligation.

The plastic correction by tantalum cranioplasty for congenital absence *of portions of the skull is not recommended until the age of rapid cranial* growth has passed. Such cranial defects will not close spontaneously by bone growth and must be repaired by plating with tantalum by the age of 5 or 6 years. In extremely large defects a type of padded helmet will give added protection to the head during the time the child is awaiting plastic repair. The treatment is fully discussed under Chapter VI on plastic surgery.

Congenital Dermal Sinus Tracts

Sinus tracts of congenital origin may exist at any point along the posterior cerebrospinal axis. Resulting from an incomplete differentiation of epithelial ectoderm from the neuroectoderm they persist as small epithelial-lined tracts which extend from the surface to end at any point down to the spinal cord or brain. Usually found in the lumbosacral and occipital areas they are detected by the presence of an innocuous appearing dimple in which a tiny, patent opening is found. As in spina bifida and diastematomyelia, the surrounding skin may have an abnormal covering of hair or be discolored by a "port-wine" angiomatous stain. A mass beneath the skin representing an epidermoid or dermoid cyst may be palpated, or, at times, the fibrous cord of the sinus tract may be distinguished by careful palpation. Where infection of the tract has occurred the skin about the opening may be reddened, indurated, swollen and tender. The common

pilonidal sinus represents one form of congenital dermal sinus which is frequently recognized because of recurrent episodes of infection.

The tract may terminate as an epidural, intradural, or intramedullary epidermoid or dermoid cyst depending upon the nature of the cellular elements in the cyst. Because of such a congenital portal of entry, bacterial infection may ensue, producing recurring bouts of meningitis, epidural abscess or brain abscess.

The diagnosis may be aided by the use of x-rays where a refective lamina or small occipital skull defect may be detected. Except in instances of an expanding intraspinal or intracranial cyst, the use of myelography or ventriculography is not indicated since the presence of the spinal or occipital sinus is sufficient justification for its radical surgical removal.

The operative approach to the congenital dermal sinus whether of spinal



FIGURE 19. Congenital failure of segmentation of vertebra, commonly termed "congenital fusion."

or cranial variety is to be considered a major operative procedure. Through a formal laminectomy, or suboccipital craniectomy exposure, as the case might be, the sinus tract is carefully dissected free and carried to its termination. Removal of adjacent lamina or cranium is necessary to improve the exposure if the terminus is subdural. Whenever possible the cyst is removed along with the entire sinus tract and the ellipse of skin at the distal end. However, where recurrent infection has caused dense, inflammatory adhesions about the periphery of the cyst it is wise to leave the



FIGURE 20. Hemivertebra, multiple.

innermost portion of the cyst wall where it is most adherent to the parenchymatous tissues of the cauda equina, spinal cord, or subtentorial structures rather than risk severe damage to neural tissues.

More often the tract will end blindly in the dura or pia-arachnoid and not penetrate into neural tissues. The dura and the remainder of the wound should be closed tightly in layers. The use of antibiotics before and after operation is recommended as an added protection against infection. No surgery is indicated until all evidences of clinical infection have subsided

Klippel-Feil Syndrome

Reduction in the number of the cervical vertebra associated with a failure of segmentation of the vertebrae ("fusion" of vertebrae) is termed the Klippel-Feil syndrome. Additional defects such as hemivertebra, or partial absence of a vertebra, deformity of ribs, scoliosis, kyphosis, and platybasia may be associated. Congenital elevation of the scapula



FIGURE 21. Hemivertebra, cervical.

(Sprengel's deformity) is often present. The condition is noted at birth and is characterized by an apparent absence of a neck, the head appearing to rest on the shoulders, with marked limitation of movements of the head and neck. There is no neurologic disability resulting directly from the



FIGURE 22. Severe congenital defect in cervical vertebrae producing clinical picture simulating Klippel-Feil Syndrome.

bony lesion although weakness of the arms may be present as well as clinical evidence of syringomyelia. There is no treatment.

A somewhat similar condition may involve absence of the sacral and coccygeal vertebra and is associated with defective development of the lower segments of the spinal cord. The lowest ribs may reach the crest of the ilium while the lumbar spine is quite low in relation to the pelvis. Weakness and atrophy of the gluteal muscles as well as loss of sphincter control are responsible for the term crurovesical-gluteal dystrophy used in connection with the spinal anomaly.

Basilar Impression — Platybasia

Definition and Pathology

Basilar impression is a term used to denote a disturbance in the relationship of the base of the skull to the upper cervical spine, wherein there is an invagination of the margins of the foramen magnum and the upper cervical spine into the posterior fossa. The term platybasia is used synonymously with basilar impression, but properly it is an anthropological term indicating a flattening of the base of the skull and an increase of the basal angle to more than 150°. As a result of the deformity there occurs a stenosis of the foramen magnum, diminution in the size of the posterior fossa, approximation of the arch of the atlas and the occipital bone, and stricturing of the dura at the level of the foramen magnum. The adontoid may project into the upper spinal canal. Cerebellar tonsilar herniation may occur producing an obstructive hydrocephalus. Due to the distortion of structures already mentioned, various disorders of the cranial nerves may ensue.

Etiology

The cause is unknown, but is generally considered to represent a congenital structural malformation when present in the younger age groups, at times occurring in association with other deformities such as the Klippel-Feil syndrome. Attempts to relate the disorder to rickets, osteomalacia and hyperparathyroidism have been made. In the adult, the relationship to Paget's disease is well established. There is some merit to the theory that basilar impression represents a structural weakness in the base of the skull, the actual deformity occurring slowly and progressively after erect posture has been assumed with the weight of the head in the vertical position causing the impression of the base.

Clinical Manifestations and Diagnosis

Basilar impression may be found during routine x-ray studies of the skull or cervical spine and be free of any clinical manifestations. In advanced cases the vertical diameter of the head may be shortened and the neck may



FIGURE 23. Typical radiographic appearance of platybasia.

appear shorter than normal. A veritable galaxy of neurologic signs and symptoms may be present including cerebellar ataxia, cranial nerve palsies, (especially abducens palsy) nystagmus, weakness and numbness of limbs, headache, cervical pain, papilledema and slowness of respiratory rate. The Queckenstedt test may reveal a manometric block, but in the presence of intracranial hypertension, papilledema or bradypnea a lumbar puncture is contraindicated. While these clinical manifestations may be helpful in suggesting the presence of basilar impression, the diagnosis can be established with certainty only by adequate x-ray examination. Lateral views of the skull will demonstrate the adontoid projecting above Chamberlain's line (from the hard palate to the posterior border of the foramen magnum).

Normally, this line lies above all portions of the atlas and axis. Laminagraphy may be helpful in delineating the basilar structures. In the antero-posterior projection the floor of the middle fossa and the petrous pyramids may be tilted upwards and there may be a recognizable stenosis of the foramen magnum.

Treatment

The treatment is surgical and consists of the subperiosteal exposure of the suboccipital area and the spinous processes and laminae of the upper

three cervical vertebra. The operation is carried out in the classical face-down cerebellar position under endotracheal anesthesia using a linear midline incision. In older patients the operation may be satisfactorily performed in the forward sitting position. The removal of the impressed occipital bone at the foramen magnum may be difficult until the upper cervical laminectomy is completed. A generous suboccipital craniectomy can then be completed, exposing the dura over the lower midline cerebellar area. The dura is then opened beginning with a V-flap on either side of the midline over the cerebellum converging toward the midline at the level of the margin of the foramen magnum. The occipital sinus is secured between silver clips or ligatures and the dural opening then carried well down into the spinal canal. Adhesions between the pia-arachnoid and dura are carefully freed and if there is herniation of the cerebellar tonsils they are gently lifted, separated, and freed to establish a free flow of cerebrospinal fluid. In rare instances amputation of the tonsillar herniation may be necessary. The dura is allowed to remain open. Meticulous closure of the muscles, subcutaneous tissues and skin in separate layers is imperative if one is to avoid a possible cerebrospinal fistula. Stabilization of the occipitocervical region by bone grafting is not considered necessary.

Craniostenosis

One of the encouraging chapters in the development of contemporary pediatric-neurological surgery has been the perfection of a suitable operation for the correction of craniostenosis. Long recognized as a congenital abnormality of growth, attempts to relieve it were almost universally disappointing. Currently, little is known relative to its cause, although syphilis, inflammations, and intrauterine pressures have been indicted. Most likely, the theory of an existing defective mesenchyme incapable of inhibiting ossification, is the most acceptable.

Developmentally, the cranium ossifies in its basilar portions from ossification centers in cartilage, becoming complete at about the seventh year. The vault ossifies from membrane whose margins remain unfused but in close apposition until growth is completed. The posterior fontanel is normally closed by the second or third month, the anterior by the sixteenth. By the end of the first year the sutures have become serrated and show interlocking, but actual closure by ossification is not complete until long after growth has ceased. Since 50% of all growth of the skull occurs during the first year of life it is obvious that any factor which limits growth of the cranium similarly will prevent the normal growth and expansion of the brain. Since the disease involves only the skull the nervous system is not affected except by the restriction of growth imposed by the fused sutures. The importance of early recognition of craniostenosis and the

institution of prompt surgical treatment cannot be overemphasized. Due to the peculiar, misshapen head and mental retardation in delayed cases, it may be confused with microcephaly, a condition producing mental deficiency and early fusion of the sutures due to primary failure of the brain to grow.

Diagnosis

Craniostenosis is to be suspected in any infant possessing a grossly misshapen head or premature closure of the fontanel with asymmetrical growth of the cranium. When the condition is well established, visible or palpable ridges of bone are found overlying the site of the closed suture. The condition may be obvious at birth or appear at any time during the period of active growth and may be associated with other defects such as cleft palate, high arching of the palate, syndactylism, spina bifida and meningocele. In advanced cases mental retardation may result from the mechanical arrest of normal brain growth, as well as signs of chronic increased intracranial pressure (papilledema, optic atrophy).

Depending on the duration of the synostosis and the individual sutures involved, other characteristic clinical patterns may be observed. Since longitudinal growth of the head occurs by virtue of the coronal (and



FIGURE 24 Craniostenosis involving the coronal suture. Note asymmetry of head and prominence of zygoma.

lambdoidal) sutures, premature closure will result in a head that is shorter in the anteroposterior direction and much increased in the transverse diameter with the frontal area increased in height and width *(brachycephaly)*. Fusion of a single coronal suture produces a disfiguring asymmetry of the frontal area with flattening of the frontal boss, a poorly defined orbital ridge and marked prominence of the zygomatic-temporal area.

Growth of the cranium in width depends on the integrity of the sagittal suture. Synostosis involving the sagittal suture results in a skull which is characteristically long and narrow *(scaphocephaly)*.

Premature closure of all the sutures results in a small, pointed head with prominence of the eyes due to shallowness of the orbits *(oxycephaly)*. In such children visual disturbances and mental retardation may be most severe.

Various combinations of sutures may be involved in craniostenosis but identification of the abnormal sutures involved is finally dependent on x-ray studies.

The diagnosis of craniostenosis is accurately confirmed by x-ray and



FIGURE 25. Coronal craniostenosis (Brachycephaly)

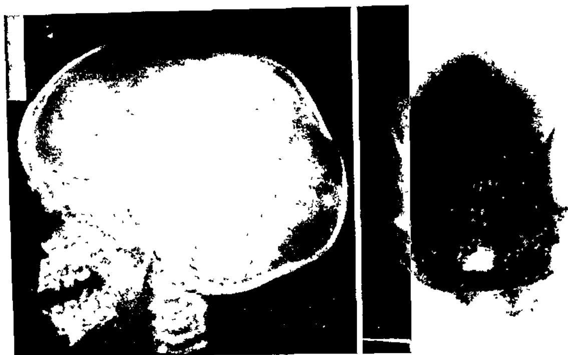


FIGURE 26. Premature closure of sagittal suture (Scaphocephaly).

will indicate the degree and extent of suture involvement. Open sutures are readily recognized and sutures that are prematurely closed will be absent or reveal their location by the presence of a ridge of bone along the site of fusion. Marked evidence of convolutional atrophy of the inner table is often observed in severe cases.

Operation

The only treatment is surgical and is indicated without unreasonable delay once the diagnosis is established. While it is true that a certain amount of cosmetic improvement occurs in the early treated cases, the primary reason for surgical treatment is to allow additional room for intracranial expansion. It is important that families understand at the outset the reason for the operation so that they will not expect an immediate cosmetic miracle.

The operation is performed under general endotracheal anesthesia with an indwelling intravenous canula or plastic catheter for blood replacement. For the sagittal suture a linear incision is carried from the frontal region to the posterior parietal. The pericranium is excised over the midline vertex area to include a strip about two inches in width. Parallel linear craniectomy is performed on either side of the midline, or a wider strip of bone can be removed from the mid-vertex area without harm, beginning just anterior to the coronal suture and ending posterior to the lambdoidal. With a cranial punch multiple openings are made about 2 cm. apart along each bony margin. A cuff of folded polyethylene film is placed along

each bone edge and held in place by interrupted sutures passed through the punch openings, thus serving as an interposing membrane the function of which is to delay the rejoining of the bone edges by new bone growth during a period in which growth of the brain is at its peak. Ultimate synostosis is inevitable, but the delaying action of the plastic film is usually sufficient to insure a gratifying degree of growth in a direction previously impossible. If union of the bony edges should occur too promptly the operative procedure can be repeated.

Craniostenosis involving the coronal suture is treated in a similar fashion, using a coronal incision from zygoma to zygoma. The craniectomy includes removal of bone well below the level of the squamosal suture bilaterally. Where coronal craniostenosis is associated with extreme shallowness of the orbits, exophthalmus and lateral deviation of the globes, decompression of the orbit must be considered. This is accomplished by extending the coronal craniectomy to include a generous excision of the squamosa to afford exposure of the orbital roof which is then removed piecemeal. The orbital decompression is carried to the lateralmost confines of the orbit. If the sphenoid ridge is unduly prominent it is also removed. If bilateral decompression of the orbit is necessary, the opposite side is treated in a similar manner when the original site has healed.

Operation for the relief of multiple suture closure involving the sagittal and coronal (and lambdoidal occasionally) are best performed through a coronal incision as described above, but placed slightly more posteriorly. By generous reflection of the scalp forward and backward sufficient exposure is obtained to allow for coronal and anterior sagittal craniectomy in a first stage operation. Later, using the same incision, a posterior sagittal and lambdoidal craniectomy may be carried out. Where indicated squamosal craniectomy can be performed, connecting the coronal and lambdoidal defects laterally and inferiorly. Exposure of the necessary areas in the oxycephalic cranium through a single coronal incision is facilitated by the natural shape of the skull and by the laxity of the scalp in young infants; however, the use of separate incisions may be necessary and when used must be separated sufficiently to avoid serious interference with blood supply to the intervening scalp.

Due to the relatively large surface areas exposed in such cranial operations, the looseness of the scalp and the vascularity of the bone and dura, blood loss is inevitable and should be replaced during the operation. A more potent danger, however, lies in the insidious loss of blood beneath the scalp in the postoperative period and may assume serious proportions if not corrected by replacement. Pressure dressings over the cranium cannot be used with safety to obviate fluid collection beneath the scalp. Such collections are removed by sterile aspiration. Skin sutures must be

placed sufficiently far apart and tied loosely enough to avoid ischemia of the intervening scalp edges.

Agenesis of the Corpus Callosum

The corpus callosum is formed during the third and fourth months of development from an outgrowth of the lamina terminalis. Rarely the corpus callosum may be completely absent, but most often the defect is one of a partial agenesis involving either the genu or the splenium. Early attempts to relate defects in the corpus callosum to a specific neurologic syndrome have not met with uniform success, so that the diagnosis of the condition was impossible except by autopsy. Since the malformation is associated with a characteristic pneumoencephalographic picture, detection of the condition is now relatively frequent. The principal distinguishing features consist of marked separation of the lateral ventricles, bicornuate shape of the bodies of the lateral ventricles with concavity of the medial borders, and a dilated third ventricle rising higher than usual.

In most instances the disorder is found in the course of a neurologic investigation for epilepsy or mental retardation although the condition may be present in the healthy individual without neurological symptoms. Recognition of the defect is important clinically since it must be differentiated from the ventriculographic picture produced by a neoplasm of the corpus callosum. Occasionally agenesis of the corpus callosum is associated with a lipoma in the region of the callosum.

Microcephaly

This congenital abnormality refers to a defect of development characterized by mental deficiency, a small head, early closure of the fontanels and retardation of progressive growth of the head. It is to be differentiated from craniostenosis with which it might be confused. In microcephalic children the disproportion between the size of the head and body is obvious at birth and such a disproportion remains throughout life, although there is slow growth of the head during the normal growth period. The appearance of the head is quite characteristic, revealing a generally small head which is narrow, flattened over the occiput, and with a sharply receding forehead, the effect giving a pointed appearance to the vertex. The basic abnormality, however, lies in the defective brain rather than in the skull. Except for domiciliary care and training within the potential capacities of the child, there is no treatment

Anencephaly

Such a monstrous failure of central nervous system development is included here to emphasize the degree that developmental defect may

or non-communicating. It is generally agreed that most of the fluid is formed from the choroid plexus and that some comes directly from the walls of the ventricles. The question of whether the choroid plexus acts as a filter or as a secretory body is still unsettled.

The cerebrospinal fluid circulates from the lateral ventricles through the foramina of Monro into the Third ventricle, through the aqueduct of Sylvius into the Fourth ventricle leaving through the foramen of Luschka and Magendie to flow into the subarachnoid space over the cerebral

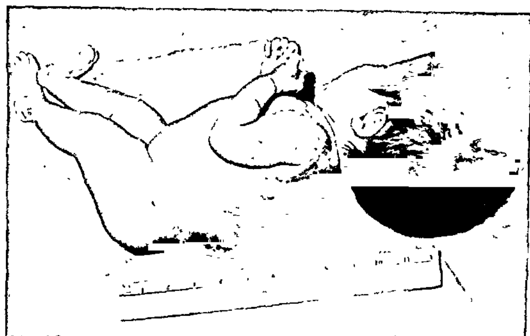


FIGURE 1. Hydrocephalus associated with a lumbar myelomeningocele

hemispheres and around the spinal cord. The great majority of fluid is probably absorbed through the arachnoid villi and perivascular spaces over the hemispheres back into the blood stream.

Over 99% of all cases of hydrocephalus are due to some blockage to the flow or absorption of the fluid. There have been several cases accurately recorded in which there appeared to be an overproduction of fluid. These were infants with papillomas of the choroid plexus in which the ventricular system was enlarged and under increased intracranial pressure but with no evidence of a block to the circulation or impaired absorption.

The etiology of hydrocephalus is due either to congenital malformations, scarring from inflammatory or traumatic processes, or neoplasms. Most of these conditions are discussed more appropriately in their respective chapters elsewhere in this book. The most frequent cause in the communicating type is some defective mechanism in the absorption of the fluid over the hemispheres. If the hydrocephalus appears within the first month or two of life, there is probably some congenital malformation in

the absorbing mechanism. However, if the situation occurs later on in childhood, it probably represents the scarring process affecting the arachnoid villi or the veins themselves as a result of some earlier inflammatory or traumatic episode. A basilar meningitis may leave the subarachnoid spaces beyond the foramen of Luschka obliterated so that the flow of fluid is blocked and unable to reach the normal areas of absorption. Consequently a communicating hydrocephalus results. Any blockage within the ventricular system will lead to a non-communicating type. The most common cause in the group under discussion is involvement of the aqueduct of Sylvius. Forking or stenosis of the aqueduct produces a hydrocephalus at birth or even in utero. Gliosis of the aqueduct more often occurs in childhood and early teens. This often is a mild inflammatory reaction of the subependymal layer. Glial septa which obliterate the foramen of Luschka and Magendie cause the fluid to block at the exit of the fourth ventricle, resulting in a non-communicating hydrocephalus detected shortly after birth. The hydrocephalus associated with myelomeningoceles is non-communicating due to the downward herniation of the cerebellar tonsils and medulla into the foramen magnum, thereby obliterating the foramen of Luschka and Magendie. Only about 10% of patients with meningoceles develop hydocephalus though frequently accelerated after the plastic repair of the spina bifida.

Signs and Symptoms

The diagnosis of hydrocephalus in infants is usually made without difficulty because of the obvious cranial enlargement and bulging of the anterior fontanelle which are indicative of increased intracranial pressure. The enlargement of the head is confirmed by measuring the greatest occipito-bregmatic circumference. The normal values are shown in Figure 2. In early cases with borderline enlargement repeated measurements at bi-weekly intervals will disclose an abnormally rapid growth. The enlarge-

Age	*Boys*	*Girls*
1 mo.	35.75-38.75 cm.	35.1-37.9 cm.
3 mo.	39.25-42.1	38.4-41.2
6 mo.	42 2 -45.1	41.1-43.9
9 mo.	44.3 -47.1	43.2-46
1 yr.	45.4 -48.2	44.4-46.8
1½ yrs.	46.5 -49 3	45.7-48.3
2 yrs.	47.6 -50.6	46.7-49.3
2½ yrs.	48.4 -51.2	47.5-50.2
3 yrs.	49.0 -51.75	48.2-50.9

FIGURE 2. Range of normal head circumference for boys and girls from one month to three years.

ment is limited only to the cranial portion of the skull and the disproportion between the large cranium and the normal face may be quite striking. The measurements should be carried out by the same individual if possible and with the use of a centimeter rule. The centimeter rule has the advantage over an inch rule in that increases of smaller units will be more easily recognized.

The enlargement of the head varies from one that is hardly recognizable to one so large that the infant, unable to turn the head from either lateral position, frequently develops pressure areas of skin necrosis on the scalp. As the head begins to enlarge and the intracranial pressure becomes elevated, the scalp veins become distended, more prominent, and tortuous. The anterior fontanelle is enlarged ranging from slight to extensive depending again on the degree of hydrocephalus. It is also bulging and tense. At times it may be extremely tight, which would mean that the intracranial pressure is high and that the clinical situation is acute requiring emergency measures for the reduction of pressure. In the very young infants the posterior fontanelle may still be open and tight. When palpating the anterior fontanelle in an early case, the infant should be held in an upright position and given a bottle so that he will not cry. Even in a normal infant, the anterior fontanelle will appear full and raised when crying in the upright position.



FIGURE 3. (*Left*) Early hydrocephalus with tight, enlarged anterior fontanella. (*Right*) More advanced hydrocephalus.

As the hydrocephalus progresses, the cranial sutures become separated, and can easily be palpated. Percussion of the head reveals a "crack-pot" sound due to the increased amount of fluid and smaller amount of brain tissue. The eyeballs may be displaced downward by pressure upon the orbital plate so that the sclera is visible above the iris. The extraocular movements of the eyes are incongruous and one eye will move independently of the other eye. The pupils are most often equal, though dilated. Optic atrophy takes place in the late stages. The patient becomes more irritable and develops a high pitched cry as the enlargement progresses, especially when the anterior fontanelle becomes very tight. Feeding is no problem until the intracranial pressure becomes excessively high. However, the hydrocephalic infant does not eat quite as well as the normal infant and consequently his body appears somewhat undernourished. The disproportion between his head and his entire body becomes quite pronounced even to the inexperienced examiner.

When the anterior fontanelle becomes very tight, the infant may appear lethargic and begin to vomit. Soon thereafter, depression of the vital functions occurs and death intervenes. However, in the very young infant death does not take place until many months or even several years after the onset. Often the cerebral cortex will be only millimeters thick and yet vital functions are not impaired.

Mental retardation is the symptom most often feared should the baby survive. There is no direct correlation between the thinness of the cerebral cortex and the degree of mental retardation. If the hydrocephalus occurs slowly over a long period of time, the amount of mental retardation will not be as pronounced as if the hydrocephalus took place over a short period of time. Nevertheless, mental retardation of varying degrees occurs whenever the hydrocephalus has been present for any length of time. It is certainly possible that when the hydrocephalus has been present for only a short period of time and subsides or is successfully surgically corrected, mental retardation may not occur.

The hydrocephalus may become arrested when spontaneous absorption of the fluid takes place. This may occur several weeks to many months after the onset of the hydrocephalus. When this occurs the size of the head no longer increases and the anterior fontanelle remains wide but is no longer tight and bulging. However, the mental retardation that has already taken place does not recede.

The signs and symptoms of hydrocephalus occurring at an age after the cranial sutures are closed are those of acute increased intracranial pressure and the head size does not increase with the progressive enlargement of the ventricles. These symptoms are described under Chapter XII on brain tumors, but briefly consist of headache, diplopia, nausea, vomiting, drowsiness, papilledema, and extraocular palsies. A critical picture develops

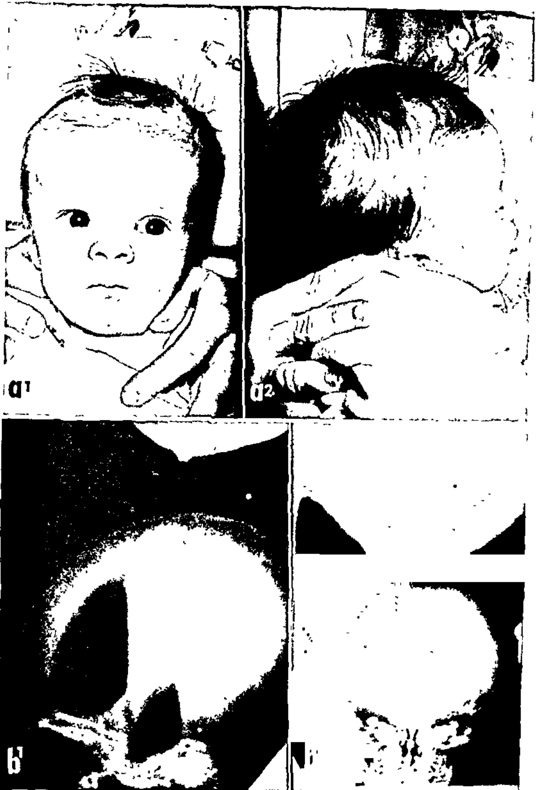


FIGURE 4. Early spontaneous arrest of hydrocephalus. (Hydrocephalus probably developed in utero and spontaneously arrested there before delivery.) (a) Frontal and lateral view of a one month old child born with a large head which did not continue to increase at an abnormal rate. The anterior fontanelle was enlarged, but soft and depressed (b) Brow up lateral and AP view of ventriculogram taken on same infant a day after photograph in Figure 4a was made. Note tremendous size of the ventricular system yet there was no increase in pressure nor did the head continue to grow at an abnormal rate in subsequent years.

much more rapidly since the cranial vault does not enlarge along with the increase in intracranial pressure. The depression of vital signs such as slowing of the pulse and respiratory rate appear within weeks to a few months after the onset and necessitate early surgical intervention. The patient's neck may become slightly stiff on flexion, which usually means that the cerebellar tonsils are herniating.

Diagnostic Procedures

X-rays of the skull are routinely done. They will show varying degrees of suture separation and thinness of the calvarium. The roentgenograms merely confirm the clinical impression of increased intracranial pressure and hydrocephalus. Occasionally, abnormal calcifications may be seen when a neoplasm or subdural hematoma is present.



FIGURE 5. (a) Lateral view of the skull in hydrocephalus revealing the markedly separated sutures, and a disproportionate cranial-facial ratio. (b) Similar view of x-ray of skull in same patient as in Figure 5a, taken one year later following a lumbar peritoneal shunt. Note the absence of any sutural separation and no further enlargement of head. There is much less disproportionate ratio between calvarium and facial bones now than in Figure 5a.

Electroencephalography in the young infant is unnecessary and of no value. In the older child it may be done disclosing a generalized dysrhythmia but will have no localizing or diagnostic meaning.

Air contrast studies as described in Chapter II, are absolutely necessary diagnostic procedures and are carried out in the infant without anesthesia but in the older child under general anesthesia. *Ventriculography* is preferred by most neurosurgeons, but some perform *pneumoencephalography* in the infant with the enlarged head. In the older child with papilledema,

ventriculography is almost universally done through burr holes or twist drill holes. Either the bubble technique, or an exchange with 100 to 200 cc. of air is performed in the infant. The air is introduced through the lateral angle of the anterior fontanelle. After the needle has just punctured the dura the stylet is withdrawn in order to rule out any subdural collections of fluid as the cause of the hydrocephalus. Total replacement of ventricular fluid is not necessary in the infant and carries a high morbidity if not an occasional mortality. The purpose of the air study is to determine where and if an obstruction to the flow of cerebrospinal fluid exists. It is quite



FIGURE 6. Percutaneous ventriculogram showing dilated ventricles in the AP view (a), and lateral view (b). These ventricles contain over 120 cc. of air.

helpful to take some films with the baby in the upside-down position in order to get the air into the aqueduct and fourth ventricle. Usually if one sees air in the fourth ventricle and cisterna magna following a ventriculogram, a diagnosis of communicating hydrocephalus is made. This diagnosis is most often correct but for the one exception of occlusion of the foramen of Luschka and Magendie which is a rare condition having been seen four times in the past eight years at the University of Texas. This condition can be established or rejected with the addition of air introduced from the lumbar route in which the x-rays will reveal a septum between the air in the cisterna magna and in the upper cervical area, or the absence of a dye in the lumbar spinal fluid twenty minutes after it has been introduced into the lateral ventricle (dye technique discussed below). If no air is visualized below the posterior portion of the third ventricle, one may assume that the obstruction is in the aqueduct of Sylvius which would mean a stenosis in infants or a gliosis in older children. When air adequately outlines all the ventricles and cisterna magna but none over the cerebral hemispheres or basal cisterns, the diagnosis of a communicating hydrocephalus is made with the obstruction occurring at

the base of the brain. However, if the air is seen in the ventricles and basal cisterns, the block is in the cerebral subarachnoid spaces. If air is visualized in addition over the cerebral hemispheres, one may assume that there is defect in the mechanism of absorption in the arachnoidal villi or perivascular spaces near the sagittal sinus.

The *Dye test* is a method of determining whether or not the hydrocephalus is communicating or non-communicating. The dye, indigo carmine or PSP, is injected into the lateral ventricle and twenty minutes later a lumbar puncture is performed. If dye is recovered within twenty minutes, a communicating type of hydrocephalus exists. The dye may not get to the lumbar subarachnoid fluid for one to twenty-four hours or more in non-communicating hydrocephalus indicating the degree of non-communication. The dye test can be used on a patient of any age. The concentration of the dye recovered in twenty minutes is of importance. A heavy concentration is indicative of complete communication, while weak concentrations reveal some mild degree of obstruction between the lateral ventricles and lumbar fluid.

At times *Pantopaque ventriculography* under local anesthesia is performed by some surgeons. When the aqueduct of Sylvius or the fourth ventricle is not visualized in air studies even in the upside-down position in the infant, 1-1½ cc of pantopaque is introduced into the lateral ventricle under fluoroscopic control and the patient's head is manipulated so that the positive contrast material will flow into the third ventricle, aqueduct, fourth ventricle and cisterna magna unless there is an obstruction.

Upon the completion of one or a combination of these diagnostic contrast procedures, the site of the lesion responsible for the development of the hydrocephalus has been determined in most instances so that the proper surgical procedure can be carried out. If a neoplasm, subdural hematoma or effusion, abscess, or any other space occupying lesion is discovered, direct surgical intervention is performed if possible.

Treatment

Almost all of the "new" procedures for the treatment of hydrocephalus that have been introduced within the past ten years had been reported in the literature about fifty years ago. The improvement in today's results has been largely due to greater technical skill, improved materials and facilities, sterility and antibiotics, and a large number of qualified, interested surgeons, rather than any new methods for providing adequate absorption or changes in the basic physiology of the cerebrospinal fluid. The treatment is essentially a mechanical one and not one of altering the physiology. Some of the procedures are applicable for both the communicating and non-communicating variety of hydrocephalus, while others are just for

either one or the other type. The techniques described and discussed below are those that have had some measure of success with neurosurgeons throughout the United States and Canada, and have been in use long enough to be fully evaluated. However, one new technique will also be described, because it seems to show great promise even though only a few surgeons have employed this procedure with a relatively short follow-up.

Choroid plexectomy is the operation designed to decrease the amount of cerebrospinal fluid produced from the ventricles. Dandy proposed such an operation as early as 1918, in which he removed the choroid plexus after the ventricles were completely emptied of their fluid. Three of his four patients died. Putnam, in 1934, and also Scarff recorded their experiences with the endoscopic cauterization of the plexus with very encouraging results. However, this particular technique was changed to a more open operation because of the numerous technical problems encountered with the endoscopic one.

Choroid plexectomy is now performed in the following manner. A 5 cm. incision is made on one side of the scalp in the posterior temporal-parietal region with the patient under endotracheal anesthesia. The head is in the lateral position with the face turned downwards. A 1½″ opening is made in the skull with a burr and rongeurs, followed by opening the dura in a circular fashion effecting a flap. The ventricle is punctured with a needle in order to obtain the direction in which the cerebral tissue will be removed to visualize the choroid plexus. A 2½ cm. incision is made in the cortex down to the ventricle. The choroid plexus is very gently picked up with a pair of smooth tipped forceps and the base is either cauterized or divided with tantalum clips. Absolute hemostasis must be obtained. If need be, the second side can be done seven to ten days later.

Choroid plexectomy can, at the most, diminish somewhat the formation of cerebrospinal fluid since the choroid plexus is removed only from the lateral ventricles and not from the third and fourth ventricles. In 1943, Putnam recorded his results on forty-three patients operated upon between 1934 and 1942, in which eleven (25%) were hospital deaths, fifteen (33%) were later deaths, and sixteen (42%) survived. About the same time, Scarff reported his results in twenty infants in which three (15%) were hospital deaths, seven (35%) were poor results, and ten (50%) were considered good results. In ten years he recorded a follow-up of the ten patients he had described as good results. Four of the patients died of unrelated causes, three were physically well but mentally retarded, and three were receiving average or better than average grades in school. In 1952, he reported on an additional nineteen patients in which only one (5%) died in the hospital, three (15%) were poor results, and fifteen (80%) were considered good results.

These results average better than 50% relief of their hydrocephalics by choroid plexectomy. However, since the early 1950's this operation has been done rather infrequently and that would indicate that the great majority of neurosurgeons were unable to equal the results of Scarff and Putnam.

The *Ventriculo-cisternostomy* (Torkildsen procedure) is the most universally successful procedure for a *non-communicating type of hydrocephalus*. If the block is in the third ventricle, aqueduct of Sylvius, or the fourth ventricle, the Torkildsen procedure is the one of choice. This procedure has proved successful in the hands of practically all neurosurgeons. There have been many minor modifications of this procedure since Dr. Arne Torkildsen originally described it in 1939, but none of these are of any major consequence.

This operation is done preferably with the patient in the upright position but with the infant it is better to do it in the prone face down position with flexion of the neck. The incision is a occipital-suboccipital-upper cervical one in which the occipital portion is placed to the side of the midline at a distance of several centimeters. The incision is carried down to the bone which is exposed for a width of about 2″ from the occipital bone down to the foramen magnum and the arch of C-1. A burr hole is made in the occipital bone and then a channel of bone 1 cm. in width is rongeured down to the foramen magnum. One can start taking the bone off from the foramen magnum by means of a Smith-Kerrison punch. Some surgeons, including Torkildsen, do not remove this channel of bone. The arch of the atlas is removed. A ventricular needle is inserted through the burr hole into the occipital horn but as little fluid is lost as is possible. The needle is withdrawn and then a rubber or plastic catheter is inserted into the occipital horn through this needle tract, a distance of 2″-3″ within the ventricle. The catheter is then clamped off. The dura is opened over the inferior aspect of the cerebellum toward the midline taking every effort not to open the arachnoid. The arachnoid is then opened and the catheter is passed beneath the arachnoid into the cisterna magna, a distance of about 1 cm. If considerable fluid is lost from the subarachnoid space and when the catheter is placed subdurally, the arachnoid will obstruct the flow of fluid. The dural opening is closed and the catheter is anchored to prevent it from slipping.

Most reports concerning this procedure are for tumors and other causes of non-communicating hydrocephalus in adults. In 1952, Scarff reported the results in twenty patients from the New York Neurological Institute between 1941 and 1951 in which there were three (15%) hospital deaths, eight (40%) poor results in which there was operative survival without permanent relief of intracranial pressure, and nine (45%) good results.

Paine and McKissock in 1955 reported five (16%) patients as hospital deaths, nine (31%) as poor results, and sixteen (53%) as good results. Though these figures are not much better than those of the choroid plexectomy, it should be understood that actually the Torkildsen procedure has a much wider acceptance among neurosurgeons and consistently better results while the choroid plexectomy has almost been abandoned today

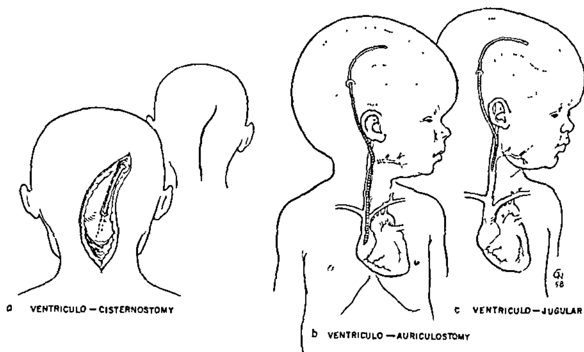


FIGURE 7. Various shunting procedures used in the treatment of hydrocephalus, I. Each procedure is adequately described in the text. (a) Ventriculo-cisternostomy (*Torkildsen's procedure*) (b) *Ventriculo-auriculostomy*. (c) *Ventriculo-jugular shunt.*

because of universally poorer results other than those presented by Scarff and Putnam.

Following the Torkildsen procedure, the patient may continue to have increased intracranial pressure even though the shunt is still functioning. This can be best established by measuring the pressure in the lateral ventricle and in the lumbar subarachnoid space simultaneously on a table in which the horizontal plane may be changed so that the head can be raised higher than the feet and vice versa. If the fluid levels in the two manometers remain in the same horizontal plane regardless of height, then one may be sure the Torkildsen tube is working even though the pressure is elevated. The explanation for this is that the cerebral subarachnoid pathways are not opened even though the shunt is working and therefore the fluid is not being absorbed. Therefore, in evaluating some of the poor results of the Torkildsen procedure this one aspect should be taken into

account. A patient still having increased intracranial pressure following the Torkildsen procedure does not mean that the poor result is due to the procedure itself. Following the Torkildsen procedure under such circumstances the patient will frequently have to have repeated lumbar punctures until the cerebral subarachnoid spaces open and the fluid is absorbed. On the whole, the Torkildsen procedure is less effective in very young infants than in older children. However, in some patients when the pressure is not decreased one may perform one of the lumbar shunts (described below). The Torkildsen procedure converts a non-communicating hydrocephalus into a communicating hydrocephalus. Therefore one then treats the patient with one of the shunts designed for the treatment of communicating hydrocephalus.

Before Arne Torkildsen introduced his operation, ***Third ventriculostomy*** was most commonly employed for obstruction of the aqueduct of Sylvius. The only indication for this procedure today would be in the patient in whom a Torkildsen procedure was indicated but rejected because of the unavailability of the cisterna magna for one reason or another. Even then it might be preferable to carry out one of the ventricular shunts as described below. In order for the third ventriculostomy to function, the basal cistern and cerebral subarachnoid pathways must be open. Dandy first carried out this operation in 1922 by establishing a communication between the third ventricle and the cisterna interpeduncularis. Stookey and Scarff, in 1936, reported on the third ventriculostomy in which they punctured the lamina terminalis and the floor of the third ventricle. In 1951, Scarff reported on thirty-four patients that he and Stookey had operated upon between 1936 and 1950 in which there were sixteen cases with aqueduct gliosis. Two (13%) of these patients died in the hospital, while four (25%) were considered poor results and ten (62%) were considered good results with a survival period ranging from six weeks to fifteen years. The operation is carried out through a small, right frontal craniotomy with elevation of the frontal lobe. The opening is made into the anterior portion of the third ventricle which is seen bulging over the chiasm. To further insure drainage, another opening is made through the posterior part of the third ventricle into the interpenduncularis cistern.

Lumbar subarachnoid ureterostomy in which the excess spinal fluid is diverted from the lumbar spinal canal to the ureter was first reported by Heile in 1925 and modified and successfully popularized by Matson since 1948. It is applicable only to the communicating type of hydrocephalus. The initial author abandoned the procedure because of a high mortality rate. Today Matson's modification is one of the more universally performed techniques in the treatment for communicating hydrocephalus yielding

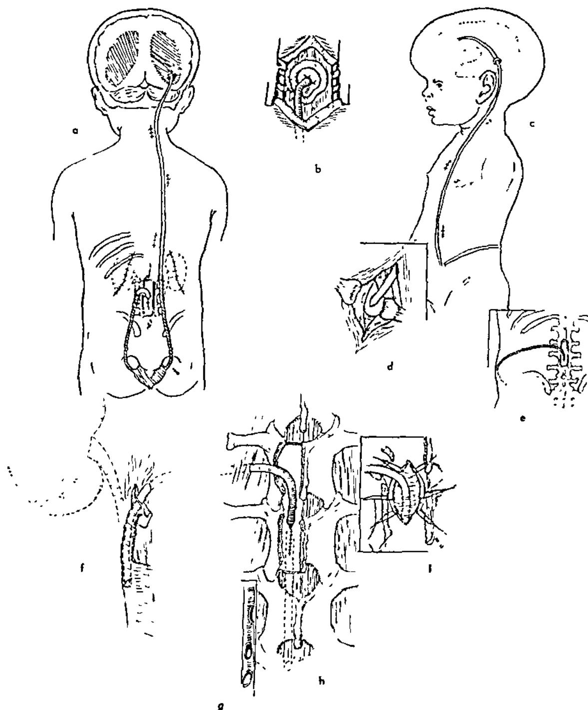


FIGURE 8 Various shunting procedures used in the treatment of hydrocephalus, II Ventricular and lumbar subarachnoid shunt to the ureter and peritoneum. (a) A ventricular-ureteral shunt (right) and a lumbar subarachnoid ureteral shunt (left). Obviously both shunts are never done on the same patient since removal of a kidney is necessary to make ureter available for each tube, unless in an unusual circumstance both tubes are inserted in the same ureter. (b) Cranial burr hole with tube inserted through the dura Though not seen, the tube penetrates the underlying piarachnoid and cerebral tissue to enter a lateral ventricle as shown in cranial portions of (a) and (c) The inferior portion of the tube is passed subcutaneously down to

fair results in the hands of many neurosurgeons. However, Matson's results are quite good in that about 70% (41 out of 58 patients) are living and asymptomatic following the operation.

The technique is carried out under general anesthesia with the patient in the lateral kidney position and a subcostal incision is followed by dividing the muscles of the flank. The false renal capsule is opened after the peritoneum has been reflected medially and then the kidney is mobilized. The ureter, after being identified, is divided as it joins the renal pelvis. The kidney is removed following the division of the vessels to the renal pedicle. Matson uses the left kidney because it is somewhat higher and on the side of the aorta rather than the vena cava. A small midline, upper lumbar incision is made followed by a laminectomy of L-2 and 3. The dura is opened in the midline without opening the arachnoid. A pinpoint hole is made in the arachnoid and a polyethylene tube is inserted through the arachnoid and passed caudally for about 4 to 5 centimeters so that it floats freely among the spinal roots. The dura is closed and the tube is passed through the paraspinal muscles into the perinephric space and 4 to 6 centimeters into the lumen of the ureter.

The most important postoperative problem and the major cause of death is disturbance of fluid balance. There is continuous loss of sodium from the body through spinal fluid lost into the ureter. For this reason, vomiting, excessive perspiration, diarrhea, or lack of normal intake with any intercurrent infection may bring about acute dehydration if additional salt is not given.

Up through 1954, Matson had operated upon fifty-eight patients of whom forty-one were living and asymptomatic. In an analysis of forty-seven patients, eleven died of which six were due to inadequate fluid administration, five were re-operated upon because the catheter became obstructed in the interspinal portion a number of months later but when the tube was replaced the patient once again was asymptomatic. Matson felt that most of the patients he had selected would develop secondary pathways for cerebrospinal fluid circulation within time and that the shunt was needed only for a few months to a few years in order to carry

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the peritoneal cavity or ureter. (c) Ventricular and lumbar-peritoneal shunt. (d) Abdominal muscle splitting incision with tube having entered the peritoneal cavity. (e) Lumbar-peritoneal shunt. (f) Tube entering ureter after kidney has been removed. (g) Multiple side vents in last 2 cm. of tube within spinal canal. Tip of tube in ventricle, peritoneal cavity, or ureter has same appearance. (h) Hemilaminectomy (total laminectomy may be used) with tube placed caudally in subarachnoid space and dural opening sutured tight (either ureteral or peritoneal lumbar subarachnoid shunt). (i) Lumbar hemilaminectomy with dura opened and tube inserted through punctured arachnoid (and passed caudally in the subarachnoid space).

them over this period of hydrocephalus. He later reported that 65% (42 out of 64 children) pesented satisfactory results following this operation and that about 10% had to be re-operated upon because of occlusion of the subarachnoid end of the tube. Also, about 10% experienced aborted spinal fluid infections, though usually controlled by antibiotics, with the exception of three who died. About 20% of the patients in this later series eventually died of an improper electrolyte balance, mostly during periods of fever and gastrointestinal upsets at home.

Lumbar subarachnoid peritoneostomy has gained popularity and a moderate degree of success in the treatment of communicating hydrocephalus. A single such case was reported by Ferguson as early as 1898. However, the recent popularity since 1949 was due to Cone and later one of his pupils, Jackson. Though Cone has had the largest experience with this technique he has not reported any of his results which have been moderately successful. In 1955, Scott and his associates reported a group of thirty-two infants in which sixty-seven operations were done with a case mortality of 38% and the survival rate of 62% in cases of communicating hydrocephalus. However, they stated that this technique along with the ventriculo peritoneostomy (described below) has controlled hydrocephalus in but a small percentage (9%) in their cases over a period of four years. However, Jackson and Snodgrass in the same year, reporting on sixty-two patients (although some of these were adults and also included those with ventriculo peritoneostomy), had a total of 116 operations. They believed that only 39% of their patients were benefited over a long period of time by either of these techniques. The results for short periods of time are much more impressive.

The operation is carried out under general anesthesia with the patient in a lateral semiflexed position with the left side uppermost. A small mid lumbar incision is made and a hemilaminectomy is carried out with the removal of two lamina. The dura is opened taking care not to puncture the arachnoid. Then the arachnoid is opened sufficiently to permit a tube, polyethylene or rubber, to be passed caudally in the subarachnoid space.

The dura is closed and the tube is passed subcutaneously into either the left lower abdominal quadrant or to the midline just below the sternum. After the peritoneum is opened either in lower abdomen, the tube is passed distally along the anterior wall of the peritoneal cavity away from the omentum, or in upper abdomen, it is passed over the dome of the liver. In older children the same procedure is carried out except that the hemilaminectomy is omitted. Instead, a small incision of a centimeter or so is made through the skin in the mid lumbar region followed by the insertion of a Touhy needle into the subarachnoid space with the bevel pointing caudally. The stylet is withdrawn and a polyethylene tube is inserted



FIGURE 9. (a) Brow up lateral view of the ventriculogram again showing more enlargement than was apparent from the clinical evidence of hydrocephalus. (b) Postoperative photograph of patient showing depressed anterior fontanelle. (c) AP view of x-ray of abdomen postoperatively showing position of lumbo-peritoneal tube. The tube (silicon) has enough opacity to be visualized on x-ray but not enough to be reproduced and therefore was blacked in. (d) Lateral view of x-ray of lumbar spine showing position of tube as in Figure 9c.

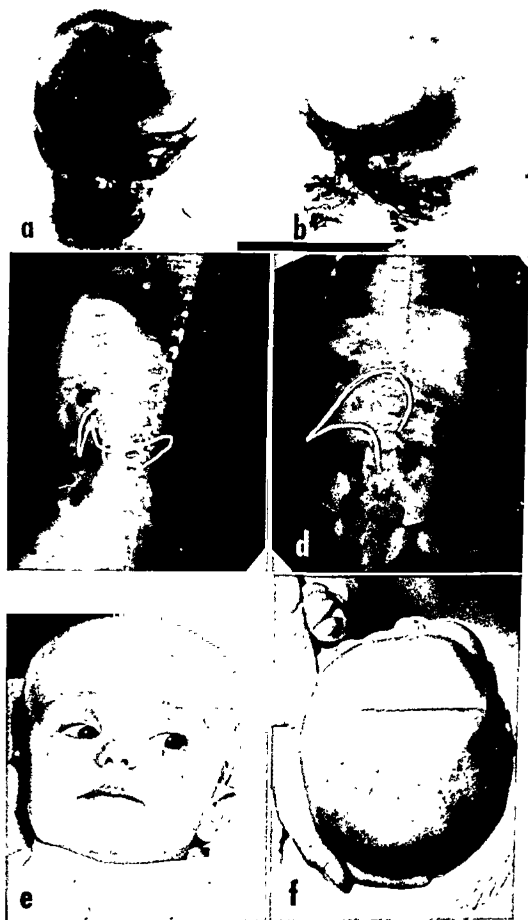


FIGURE 10

through this No. 13 Touhy needle into the lumbar subarachnoid space. The Touhy needle is then withdrawn and the other end of the tube is passed subcutaneously into the peritoneal cavity as described above.

One of the major complications of this procedure was the blocking of the tube either in the spinal canal or in the peritoneal cavity. Kinking of the tube has accounted for some obstructions. In some instances the tubes have slipped out of either the lumbar canal or the peritoneal cavity.

The best results obtained in the author's series were those in which the communicating hydrocephalus was due to some defective absorption mechanism. However three such patients developed acute increased intracranial pressure two years after operation because they had outgrown the original tube. The second tube continued to work for a period of one and one-half to two and one-half years afterwards.

There have been described some variations to this technique. Harsh put the peritoneal end of the tube into one of the Fallopian tubes. His report was of too short a follow-up to be of any value. Picaza in Havana placed the peritoneal end of the tube in the lesser omental sac with promising results.

This technique is a very simple surgical procedure and has sufficient merit to continue to be used in certain selected cases.

Ventriculo-ureterostomy is one of the two universal techniques for hydrocephalus in that it can be performed in any age patient regardless of where the block may exist, or the etiology. Up to 1954, Matson had performed this procedure eleven times in which three patients had to have the tube replaced twelve to twenty months after the initial procedure because of outgrowing the tube.

A burr hole is made in the occipital temporal region followed by one end of a polyethylene tube being inserted into the lateral ventricle, a distance of 5 cm. The other end of the tube is then passed subcutaneously down the posterior aspect of the neck and back to join the wound made for the nephrectomy as described above. After the kidney has been removed this end of the tube is inserted into the lumen of the ureter for a distance of 4 to 6 cm.

Many other surgeons have also used this technique but the results are generally less impressive than those of the lumbar subarachnoid ureter-

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FIGURE 10. (a) and (b) AP and lateral views of ventriculogram revealing tremendous enlargement of ventricles with a very thin cerebral mantle. Note the marked enlargement of the cranial mass as compared to the facial bones. This hydrocephalus occurred following a meningitis. (c) and (d) X-rays of the lumbar spine and abdomen in the AP and lateral projection showing the position of the lumbar-peritoneal tube (though visualized on x-rays it was necessary to black it in for photographs). (e) and (f) Postoperative photographs of patient revealing the enlarged but sunken anterior fontanelle indicating that the tube is functioning.

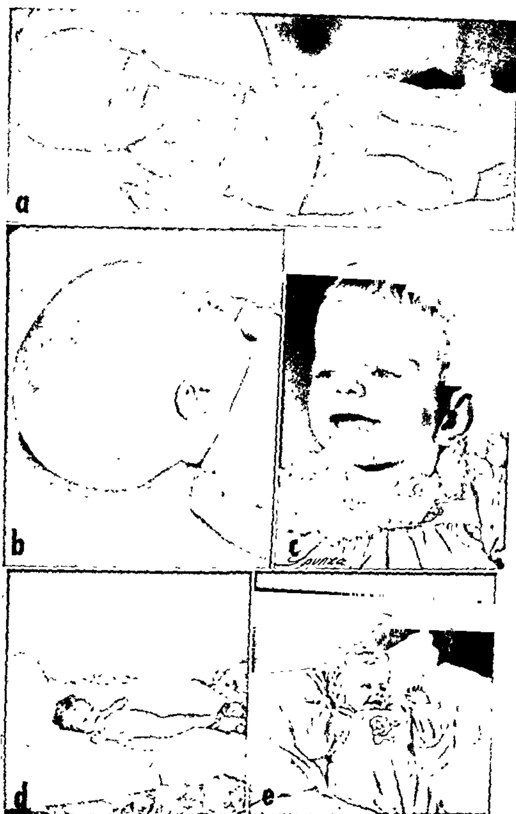


FIGURE 11. *Non-communicating* hydrocephalus *due to occlusion of foramen of* Magendie and *Luschka* by congenital septa treated successfully by *ventriculo-peritoneal* shunts after posterior fossa exploration with removal of septa *failed.* (a) and (b) Age one month. Enlarged head with tight full anterior fontanelle. (*Abdominal* bandage *represents unsuccessful* lumbar-peritoneal *shunt. Since* large *amounts of air was visualized in*

ostomy mainly because of the growth factor in the infants and in addition the same electrolyte imbalance complications that accompany the removal of a kidney with excessive sodium loss in the lumbar shunt.

Ventriculo-peritoneostomy is the other shunting procedure applicable to all types of hydrocephalus, first reported by Karsch in 1905, but nothing much was done with this technique until after World War II, when Cone performed a number of these procedures. The cranial part of this shunt is the same as that in the ventriculo-ureterostomy and the peritoneal aspect is the same as that in the lumbar subarachnoid peritoneostomy. The only difference is that the cranial burr hole is made above and behind the ear and then the tube is brought subcutaneously down the side of the neck on to the anterior chest wall entering the peritoneal cavity from the ventral aspect. The complications are the same as those in both the lumbar peritoneostomy and the ventriculo ureterostomy. On the whole, the ventriculo peritoneostomy does not work as consistently and as efficiently as the lumbar peritoneostomy.

If one is in doubt as to where the block is, either ventricular shunt would be indicated.

Nosik in 1950, advocated joining the lateral ventricle to the mastoid antrum and his early results appeared promising. However, with the longer follow-up, this procedure has failed to alleviate hydrocephalus.

Ventriculo-auriculostomy. One of the newest and yet most promising techniques is Pudenz's ventriculo-auriculostomy. The essential feature of this shunt is incorporation of a valve which reportedly prevents the tube from becoming thrombosed or blocked once the intracranial pressure has become lower than the intravascular pressure. Ventriculo-vascular shunts were first carried out about fifty years ago, but unsuccessfully. The tube worked as long as the intracranial pressure was high, in that it would prevent the back flow of blood into the tube. But once the intracranial pressure decreased, the vascular pressure would be higher and consequently blood would pass into the tube and clot, thereby obstructing the flow of spinal fluid. However with the use of the valve, which is adjusted for a certain pressure, the ventricular pressure can become low and yet no blood is allowed to flow retrograde and consequently block the tube. Nulsen and Spitz have also devised a similar valve placed in the internal jugular vein in the neck shunting the fluid from the lateral ventricle. Both of these valve procedures are also universal techniques in that they can be used for the treatment of

posterior fossa, a diagnosis of communicating hydrocephalus was incorrectly made.) (c) Age one year. Successfully managed by ventriculo-peritoneal shunts. First shunt plugged at age of six months and redone. (d) Age one and one-half years. Shunt successfully functioning (e) Age two years. Shunts successfully functioning. This is third ventriculo-peritoneal shunt.

hydrocephalus, regardless of where the block is. Spitz has performed over a hundred such procedures, but more than 50% have not been followed over six to twelve months and therefore cannot be adequately evaluated. On the other hand, Pudenz has done relatively few patients, although he had had considerable experience with this valve in animals over a long period of time. Because Pudenz's procedure seems to have greater merit among the valves used, we shall describe his technique.

The patient is operated upon lying on his back with his face turned to the left, exposing the right side of the head. An incision is made along the anterior edge of the right sternomastoid muscle and down to expose the internal jugular and common facial veins. A 3 cm. incision is then made in the right posterior temporal region followed by a burr hole and opening of the dura. A polyvinyl tube is passed through this hole into the lateral ventricle, a distance of 7 to 8 cm. This tube is then brought subcutaneously to the neck wound. The valve is attached to this end of the tube and passed through the common facial and internal jugular vein into the right auricle of the heart. The success of this technique dictates that the valve be located in the auricle so that it floats in a pool of blood. If the valve remains in the internal jugular vein, it will cease to function within a very short period of time. The disadvantage of this procedure is the same as in all shunting techniques which is the growth of the infant while the length of the tube remains unchanged. Pudenz points out that the chief advantages to this technique are the simplicity, minimal surgical trauma, adaptability to even the smallest infant, and retention of water, electrolytes and other cerebral spinal fluid constituents within the organism.

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CHAPTER X

Inflammatory Diseases

C. D. HAWKES

INFECTIONS *involving the nervous system and its coverings in childhood which require surgical treatment have become much less common since* the advent of chemotherapy. Cranial or intracranial complications of otitis media, osteomyelitis, and brain abscess all occur with much less frequency because of the eradication of the initial source of the infection by antibiotics. However, other problems have arisen which are the result of survival of children who would formerly have died from the nervous system infection they suffered. Children now recover from types of meningitis which were universally fatal in past years, but their recovery may be attended by complications such as subdural effusion or adhesive arachnoiditis and hydrocephalus. These complications following the initial infection must be treated surgically if recovery is to be satisfactory and severe residual neurological crippling avoided.

In this era of chemotherapy an important point to be remembered is that collections of pus must be evacuated. An abscess itself may well have a necrotic core to which the blood supply does not penetrate and which the chemotherapeutic agent does not reach. Even though the systemic effects of the infection are controlled, the abscess may serve as a source of further trouble and of re-infection. Surgical drainage is as important today in the treatment of such infections as it was in the pre-antibiotic era.

In addition to reducing infections of the nervous system and intracranial spread of infections about the head, chemotherapy has rendered surgical treatment more effective in those infections which do occur. Infections which formerly might have been overwhelming and fatal in outcome may now many times be brought under control by a combination of antibiotic therapy and surgical drainage. In any case where surgery is undertaken, the chance of spread of the infection is lessened by the concomitant administration of antibiotics. Thereby, more radical and definitive surgery, such as the total excision of a brain abscess, has become possible. Furthermore, primary or at least earlier closure, and consequently earlier healing and less morbidity, has been brought about.

Penicillin continues to be an effective agent against many of the organisms responsible for nervous system infection, and where it is not, broader spectrum antibiotics such as tetracycline and its modifications are likely to prove effectual. Streptomycin may be employed in tuberculous infections, often in combination with other agents, and may also be useful in combination with penicillin against certain organisms. It is important that culture material be obtained, if possible, before the institution of chemotherapy. Sensitivity tests should be carried out in order that the most effective chemotherapeutic agent can be employed. A resistant organism may be encountered in which the usual antibiotic employed for its type has little value, and testing is important to determine what drug can be used. Many different preparations of penicillin and streptomycin are being developed, and new "mycin" drugs are regularly appearing on the market. Side effects are being lessened and range of action broadened. One must therefore keep abreast of these advances in order to be aware of the proper antibiotic to employ in any case at hand, particularly in those cases where the organism is not sensitive to presently popular preparations. In 1957 and 1958 many cases of hospital infection with resistant staphylococci have been reported. These strains have apparently become resistant to the antibiotics currently used. Steps to control such infection are (1) increased vigilance in aseptic techniques both in the operating room and elsewhere, and (2) the reservation of newer antibiotics effective against these strains for cases actually becoming infected rather than employing them for prophylaxis. It is anticipated that the problem of resistant organisms will continue to exist and that the two steps outlined will be even more important in the future in the control of infections occurring in the hospital.

Chemotherapy may also be employed locally in selected cases. Wounds may be irrigated with solutions of antibiotics, or such drugs as Bacitracin used topically. Streptomycin powder may be used in the cavity of a tubercular spinal abscess following curettage.

Chemical debridement may be an aid in clearing up grossly contaminated and infected wounds. Trypsin derivatives and other enzyme derivatives such as a Streptokinase-Streptodornase preparation both help to clear away dead tissue and break down fibrous barrier zones to permit free penetration of antibiotics. These agents may also be effective when administered parenterally in cellulitis and deeper-seated abscesses.

Infections of neurosurgical interest in childhood may best be considered in relation to the anatomical structures involved, namely the scalp, the skull, the spine, the meninges, the brain, and finally the spinal cord.

Infections of the Scalp

Infections of the scalp deserve prompt attention, because they are liable

to lead to osteomyelitis of the skull or to spread intracranially by means of emissary veins passing through the skull.

Proper debridement of traumatic wounds of the scalp will prevent many



FIGURE 1. Pyogenic Abscess of the Scalp. Microscopic section of a pyogenic granuloma five millimeters in diameter removed from a scalp wound five months after it was sustained in an automobile accident. The scalp wound appeared to be healing satisfactorily when the patient left the hospital, but she was brought back four months later with the statement that there had been slight chronic drainage almost continually. Local treatment of the small granulating area failed to clear it up. No foreign body could be found. The lesion was grossly excised and the wound healed per primum. Microscopic section shows stratified squamous epithelium overlying edematous, collagenous connective tissue with a number of lymphocytes and plasma cells as well as pigmented macrophages present.

infections and greatly reduce their morbidity. Antibiotics may not be successful in preventing infection if debridement is incomplete, because the chemotherapeutic agent circulating in the blood stream may not reach the devitalized tissue. Sometimes gross infection will be overcome only to have some point of chronic drainage result from the formation of a pyogenic granuloma in a portion of the wound. Surgical excision of such a granuloma is necessary to achieve eradication of the infection and proper healing.

The topical use of antibiotics may be advisable in grossly contaminated, macerated or neglected scalp wounds, but when thorough surgical debridement and primary closure is possible, systemic chemotherapy is usually adequate. In secondarily infected wounds chemical debridement using enzyme preparations either topically or parenterally is very helpful in getting rid of devitalized tissue and breaking down the barrier to the entry of antibiotics administered systemically. However, chemical debridement can not be used as a substitute for adequate surgical drainage.

Surgical drainage should be carried out on infected hematomas of the scalp. Temporizing may well lead to osteomyelitis of the skull. Antibiotics should be administered both pre-operatively and post-operatively. In a similar fashion if antibiotics are administered in this way, infected sebaceous cysts of the scalp may be excised and primary closure of the wound done with primary healing in most cases.

Osteomyelitis of the Skull

Infection of the skull may occur as a result of contiguous spread from a suppurative process in the nasal sinuses, middle ear, or mastoid. In such cases the offending organisms are usually pneumococci or the streptococci. Hematogenous origin may occur from foci of infection in the lungs or elsewhere in the body, with the staphylococcus being the more common organism. Direct infection of the skull may likewise occur through wounds of the scalp or in operative wounds of the cranium, in which case the staphylococcus is again the more common offender, but the infection may be mixed in type. Osteomyelitis may be an acute, destructive process. However, if the infection is not fulminating, a tendency to localization and sequestration may occur. Sometimes the osteogenic property of the periosteal connective tissue is stimulated by chronic infection resulting in proliferative thickening of the bone.

Acute osteomyelitis of the skull is associated with tenderness, redness, and edema of the scalp sometimes producing a firm swelling known as Pott's puffy tumor. If the infection is severe, high fever, leukocytosis of 20,000 to 30,000 white blood cells per cubic millimeter, and prostration may occur.





FIGURE 2. *(Left)* Osteomyelitis of the Skull, Preoperative Roentgenogram. This eleven-year-old boy had a three-month history of a prominence in the left frontal region of the scalp which gradually increased in size and then began to be accompanied by headache. Shortly after onset, there was an episode of swelling of the peri-orbital tissues around the left eye, but if there was any soft tissue infection or sinusitis, it subsided without being recognized. A palpable mass in the left frontal region measuring approximately four centimeters in diameter and a smaller mass approaching the midline measuring two to three centimeters in diameter could be palpated with a definite irregularity in the skull at the borders. The roentgenogram shows irregular destructive changes giving a moth-eaten appearance in the region corresponding to the area of tumefaction The isolated area of bone in the center proved at surgery to be a sequestrum.

FIGURE 3. *(Right)* Osteomyelitis of the Skull, Postoperative Roentgenogram. The extent of the craniectomy which had to be performed to get rid of all the diseased bone is seen. Local and parenteral chemotherapy were employed and the wound drained for twenty-four hours. Healing occurred per primum, but a small area of infection appeared in the incision eight months later. This responded readily to treatment. There was no further difficulty, and tantalum cranioplasty was done to cover the bony defect twenty-two months after the initial surgery.

X-ray examination is the most satisfactory way to confirm the diagnosis of osteomyelitis of the skull. The presence of bone changes frequently lags behind the clinical picture, and *sometimes a series of x-rays over a* period of time is necessary to establish the diagnosis. In small children, particularly when there is edema of the scalp, minor degrees of bone destruction may be difficult to determine. Characteristically, there are

patches of decreased density with indistinct margins which are irregular and may contain a detached island of bone. Eosinophilic granuloma of the skull and other tumors may be difficult to distinguish from osteomyelitis by x-ray examination.

If the involvement is very early and there is no sequestration, treatment by antibiotics may be sufficient to overcome the infection. Aspiration to determine the offending organism and the proper antibiotic is desirable. However, if the destructive process is very wide with evidence of sequestration, or if a sinus tract is present, then surgical removal of the infected bone is necessary to bring about eradication of the infection. This may be done either by en bloc removal or by rongeuring away the softened involved bone back to firm bone. Chemotherapy even in these cases greatly

OSTEOMYELITIS OF SKULL

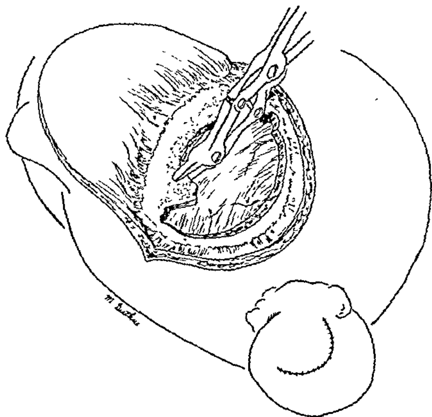


FIGURE 4. Osteomyelitis of the Skull—Operative Management. An adequate flap in the scalp must be made to fully expose all involved bone. Removal of infected bone is carried back until solid, uninvolved bone is reached. Drainage with or without antibiotic irrigation is carried out. Parenteral chemotherapy is administered.

reduces the morbidity and spread of the infection. The dura itself forms a strong barrier to spread of the infection intracranially, and prompt and adequate attack on osteomyelitis of the skull will therefore usually avoid intracranial complications.

Post-operative infection of a craniotomy wound may result in osteomyelitis of the skull. This usually involves the bone flap, which is devitalized to a greater or lesser extent (depending partly upon whether it is an osteoplastic or free flap) and provides a ready-made sequestrum. Prior to the antibiotic era, prompt removal of the bone flap was always necessary as the first step in clearing up the infection and indeed in preventing a fatal outcome from it. Even today, undue delay in removal of the bone flap, when post-operative sepsis is not brought under control promptly by chemotherapy, may result in increased severity of the infection and added risk to the patient.

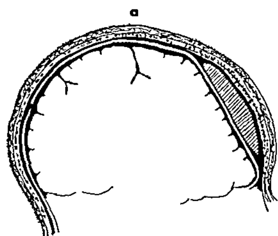
Infections of the Cerebral Meninges

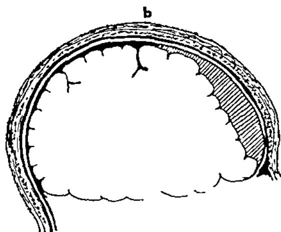
The dura mater forms a strong barrier to the passage of infection. Consequently, accumulation of pus may occur on either side of it. When this occurs between the dura and the skull, it is known as epidural abscess, and when it occurs in the potential subdural space between the dura and pia-arachnoid as subdural abscess. Such abscesses are limited in their extent, although they may spread over areas of a number of square centimeters. Infections of the leptomeninges, on the other hand, spread rapidly through the subarachnoid space borne by the cerebrospinal fluid.

Extradural Abscess

Infection in the potential space between the dura and the cranial bones is more commonly a complication of otitis and mastoiditis, but may result from any infection of the scalp or cranial bones, such as purulent nasal sinusitis, osteomyelitis or infections of the scalp. Such an *extradural abscess* (epidural abscess, epidural empyema, pachymeningitis externa) may be the result of destruction of the inner table of the skull accompanying osteomyelitis. Under these circumstances, the symptoms are tied in with those of the overlying infection in the bone, and the epidural collection of pus will be found at the time surgery is indicated for the osteomyelitis. When the abscess develops beneath an intact inner table, however, the diagnosis may be more difficult. Such an occurrence appears to result from a septic thrombophlebitis of the dural veins. There is an accumulation of pus or granulation tissue between the bone and the dura, and the dura becomes stripped away by extension of the infectious process. The development of meningeal signs fairly early in a case of frontal sinusitis should

suggest the diagnosis. Persistence of pain, fever and leukocytosis after proper treatment of other local infections around the head in the absence of osteomyelitis also should put one on the alert for this possibility. Focal neurological signs or convulsive seizures may occur if the pus accumulation becomes extensive. Tenderness over the skull in the absence of edema of the scalp or other evidence of osteomyelitis plus increasing sepsis with positive blood culture are other signs that may appear.





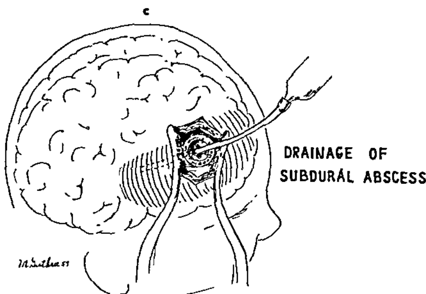


FIGURE 5. Epidural and Subdural Abscesses. Figure (a) shows the location of pus formation in extradural abscess with the accumulation lying between the skull and the dura. Subdural abscess with the pus collecting between the dura and brain is illustrated in Figure (b). The lower Figure (c) shows a subdural abscess being drained through a bur hole and opening in the dura with catheter irrigation. A drain will be left in the cavity.

Drainage of an extradural abscess usually leads to a favorable result if it is undertaken before extension of the infection to the leptomeninges and brain substance occurs. Bur hole drainage may be adequate when the abscess consists largely of free pus, but if there is granulation tissue or loculation present, then multiple bur holes or a single larger craniectomy may be necessary in order to thoroughly curette out the material from the extradural space. Drainage for twenty-four to forty-eight hours is usually adequate with the employment of supplementary chemotherapy. A tube may be left in the cavity if desired and antibiotic irrigation of the cavity carried out for a period of three to five days. Ten to 20 cc. of solution is sufficient, and it should be remembered that the solution may be quite dilute. Only a sufficient concentration is necessary to have its effect on the organisms directly encountered. In addition most antibiotics have an adverse effect on cerebral and even meningeal tissue if applied in highly concentrated form.

Acute Subdural Abscess

Abscess formation in the subdural space (pachymeningitis interna) is most commonly the result of virulent frontal sinusitis. When this occurs, the infection is spread through the posterior wall of the frontal sinus and breaks down the dural barrier. Meningeal signs may therefore appear more promptly than in extradural abscess but are not as severe as those appearing when a leptomeningitis complicates sinusitis. Instead there is more localized headache, usually accompanied by spiking fever and a high white blood count followed within a week's time by progressive hemiparesis. The patient appears more toxic than the usual case of early brain abscess.

This condition can readily lead to fatal consequences through spread of the infection into the leptomeninges or the brain itself. It is remarkable that the thin arachnoid can resist the advance of an infection of this sort as well as it does, and prompt recognition of the condition with institution of surgical drainage of the subdural space, as well as the abscessed sinus, is necessary if a favorable result is to be obtained. Drainage through the infected sinus alone is not enough, but rather a bur hole should be placed in the skull near enough to the center of the abscess to ensure its thorough evacuation. If the capsule is thick and tenacious, its total extirpation may be advisable through a small bone flap. If intensive chemotherapy and adequate surgical treatment of the infected sinus are carried out along with the cranial surgery, drainage will usually be necessary only for a few days. A catheter may be left in and irrigation carried out once or twice a day employing an antibiotic to which the organism is sensitive, as described for extradural abscess. Instillation of antibiotics is usually necessary only for a few days If there is much debris, parenteral administration of streptokinase-streptodornase may aid in clearing it up.

Surgical Complications of Meningitis: subdural effusions and post meningitic hydrocephalus

Infections of the pia-arachnoid usually spread rapidly through the neural axis because of the ready avenue provided by the cerebrospinal fluid circulating in the subarachnoid space. In certain instances, however, the infection may be of a character which forms local adhesions, and may become localized in certain areas, such as the base of the brain. The term *meningitis* is usually used rather than the more cumbersome term *leptomeningitis* in referring to infections of the leptomeninges. These infections are not usually a surgical problem in themselves but may lead to complications requiring surgical treatment.

The necessity for surgery arises principally in connection with two complications of meningitis. The first of these is the occurrence of subdural effusion accompanying and following the acute meningitis. The second is blockage of the cerebrospinal fluid pathways by adhesive inflammatory process.

Subdural Effusions:

The *H. influenzae* is the most common organism causing meningitis in small children, except when meningococcus meningitis occurs in epidemic proportions. Furthermore, subdural effusion most commonly occurs following meningitis due to the influenzal bacillus, although it may also be a sequel to meningococcus, streptococcus, and pneumococcus meningitis. The incidence is greatest in infants and the condition should be suspected when persisting fever, lethargy, convulsions, or unduly delayed recovery

SUBDURAL EFFUSION COMPLICATING MENINGITIS

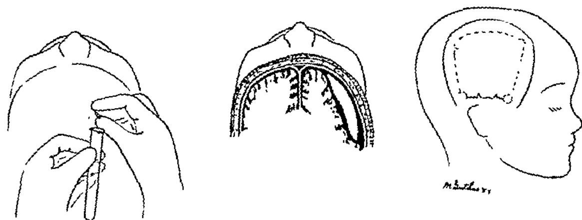


FIGURE 6 Surgical Complications of Meningitis. Subdural Effusion. Subdural taps (left) may be sufficient to relieve the condition, but if there is persisting re-accumulation of fluid and xanthochromia after four or five taps, then craniotomy (right) for excision of the membrane which forms in these cases becomes necessary. The effusion can cause considerable cerebral compression (center).

occur subsequent to an acute bacterial meningitis. A persistently bulging fontanelle may be noted in small infants. The entire problem of subdural effusions is discussed elsewhere in this volume as a separate chapter and mention is made of this occurring following meningitis.

Subdural taps will disclose an accumulation of high protein, xanthochromic fluid in the subdural space which may often have a pseudocapsule formed about it. Breakdown of the protein within the sac results in the entry of water by osmotic pressure, and by this process the subdural effusion continues to increase in size. Repeated subdural taps, daily at first and then at longer intervals, may be sufficient to clear up the condition. One may be guided by the amount of fluid obtained, increasing the interval as it becomes less. A favorable sign is decrease in the xanthochromia and therefore in the protein content of the fluid. If definite headway, both in decrease in volume of reformed fluid and in protein content, is not evident after four or five taps, bur hole drainage should be undertaken. The finding of a membrane, under these circumstances, is sufficient indication for craniotomy to effect its removal. If no significant degree of membrane formation is present, irrigation through one or more bur holes and drainage may be sufficient to stop the re-accumulation. Response of the patient is usually favorable unless the condition has gone on long enough to cause wide-spread brain damage.

Expansion of the brain may be a problem if the subdural effusion is of long-standing or is very large. Several solutions of this problem have been advocated. Intraventricular or spinal injection of saline or other physiologic solution to add to the bulk of the cerebrospinal fluid may be effective but may also cause brain damage. Prolonged external drainage of the subdural space on the other hand adds a risk of secondary infection. Subdural-pleural shunt may be more safely employed than either of these methods; the pleural cavity appears to take up high protein fluid fairly successfully. In infants and small children particularly, where the head has undergone enlargement, a plastic procedure on the dura and the skull may be undertaken to fit them to the size of the brain. Such a proceeding does not appear to interfere with later skull growth.

Post-Meningitic Hydrocephalus:

Obstruction of the aqueduct of Sylvius, the formina of Luschka and Magendie, or the basal cisterns and/or the cortical subarachnoid spaces by inflammatory adhesions may complicate meningitis with resulting hydrocephalus. This is probably a more frequent cause of hydrocephalus than is generally recognized, particularly when the initial intracranial infection may have been mild or self-limiting and masked by the empiric use of antibiotics for fever In its severer forms this condition is often associated with a good deal of brain damage, and the outcome may be unsatisfactory

even if the hydrocephalus is arrested. In milder cases some re-establishment of circulation and absorption of cerebrospinal fluid may occur, and a shunt procedure in such cases is frequently needed only for a short time. The various procedures used in the treatment of hydrocephalus are fully discussed in Chapter IX on hydrocephalus.

Tuberculous meningitis, which commonly causes thick granulations over the base of the brain, is a frequent cause of post-meningitic hydrocephalus, but acute pyogenic meningitis may also have this complication. The cerebrospinal fluid may be sterilized by antibiotic therapy but the patient's condition nevertheless worsen and the head enlarge. If the obstruction is limited to the aqueduct of Sylvius, or the outlets of the fourth ventricle, a Torkildsen operation in which a catheter is placed from the lateral ventricle to the upper spinal subarachnoid space (preferable in children to the cisterna magna) may be sufficient to relieve the hydrocephalus. If there is accompanying involvement of the absorbing pathways at the base and over the surface of the brain, then one of the various shunt procedures as described in the chapter on hydrocephalus may be required. It is important that sterilization of the fluid be obtained in order to prevent spread of the infection through the shunt; a ventriculo-jugular, -peritoneal or -ureteral shunt has to be undertaken with great precaution in this regard because of the possibility of septicemia, peritonitis, or cystitis. Many times such pronounced forms of meningitis are associated with severe brain damage and if the fluid obstruction problem is overcome and the hydrocephalus arrested, one will most often have a neurological cripple as an end result. In tuberculous meningitis, when the spinal avenue of approach becomes useless because of inflammatory adhesions, bur holes are sometimes necessary when the fontanels have closed so that ventricular tap can be done and streptomycin introduced directly into the ventricular fluid. The Torkildsen operation or shunt procedures may be an adjunct to long-term chemotherapy in these patients when complications similar to those described for acute bacterial meningitis ensue.

Brain Abscess

The control of local infections which might otherwise lead to abscess of the brain has become possible in many instances through antibiotic therapy. This has greatly reduced the incidence of brain abscess in both children and adults. In the past year or two, however, there has been some rise in incidence again due to the development of strains of organisms resistive to antibiotics generally in use. A further fact which must be borne in mind is that the wide employment of chemotherapeutic agents makes the brain abscesses that do occur more difficult to recognize. The infection may be controlled to a degree in which the systemic features are suppressed and the abscess may develop very insidiously.

Abscess of the brain may result from intracranial spread of infection from the paranasal sinuses, ear or mastoid, as a complication of infections of the scalp or skull, as the result of improperly debrided and sterilized compound cranio-cerebral wounds, and from hematogenous spread from remote sources, principally the lungs (metastatic abscess). Certain pathogeneses peculiar to childhood may occur. Thus, *paradoxical brain abscess* may result when congenital heart disease allows the passage of septic emboli from remote parts of the body into the cerebral circulation without passage through the lungs. In another special instance, persisting dermal sinuses may lead to the introduction of infection through the skull, leading to abscess formation.

In the formation of a brain abscess certain well-defined pathological stages may be recognized. First, there is an encephalitic stage during which there is no frank accumulation of pus but a generalized local or widespread inflammatory reaction. As localization of the infection occurs, a

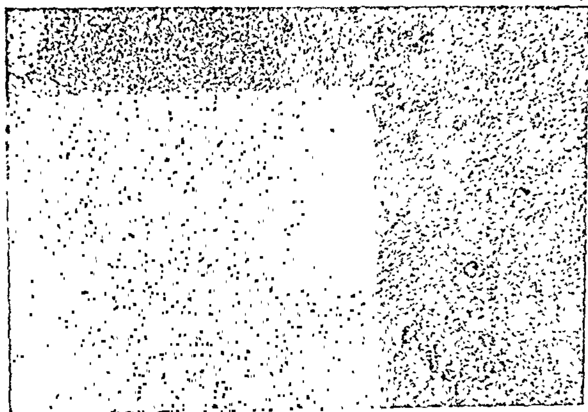


FIGURE 7. Pathology of Brain Abscess. Well formed chronic abscess occupying almost one-fourth of the cerebral hemisphere. On the left of the figure the necrotic core of the abscess containing debris and acute and chronic inflammatory cells is seen. There is next a membrane formed by fibroblasts, numerous plasma cells, lymphocytes, and eosinophiles with a few polymorphonuclear leukocytes. Within this membrane are many blood-filled capillaries. To the right of this is gliotic reaction and then edematous brain tissue.



FIGURE 8. Brain Abscess. Anterior-Posterior Roentgenogram of the Skull. Case of a two-year-old boy who bumped his head on a nail in an unfinished attic ceiling. The wound healed after a few days, but the patient later developed signs of increased intracranial pressure and a right hemisphere lesion. The small area of bony penetration in the skull to the left of the midline and separation of the sagittal suture are seen.

necrotic center develops surrounded by a leukocytic barrier. Finally encapsulation occurs from a combination of inflammatory fibrosis and gliotic reaction.

During the encephalitic stage the child will usually exhibit headache, fever, leukocytosis, stiffness of the neck and sometimes convulsive seizures. This phase may be essentially indistinguishable from a toxic encephalopathy. As encapsulation occurs, the systemic symptoms become less and the patient's temperature may often be subnormal, particularly if the acute infection of which the brain abscess is a complication has been

brought under control. The patient usually remains toxic, however, and may develop focal neurological signs depending on the location of the abscess. Localizing signs are much more common in cerebellar abscesses than those in the temporal or frontal lobes. Multiple metastatic abscesses offer a bad prognosis.

Signs of increased intracranial pressure, accompanied in infants by separation of the sutures, or the Macewen "cracked pot" sign in older children, may develop insidiously. Under these circumstances the development of increased intracranial pressure is very much like that occurring with intracranial tumor. If the initial infection which led to the brain



FIGURE 9. Brain Abscess Anterior-Posterior Roentgenogram after Ventricular Air Injection in the Case Described under Figure 8. The abscess was not encountered by the needle on the right, but very little air could be got into that ventricle. The marked degree of shift is usual in cases of brain abscess. Craniotomy was done and the abscess excised in toto with primary healing. There was minimal residual contralateral weakness.

abscess as a complication was not severe, or was remote enough so that the causal relationship is not clear, tumor may be erroneously suspected rather than abscess. Usually, however, the cerebral edema which accompanies a brain abscess leads to more rapid development of intracranial pressure and higher degrees of it than are ordinarily seen with intracranial tumor.

The brain may compensate for both the presence of an abscess and the accompanying increase in intracranial pressure for some days and then the patient may suddenly lapse into a comatose state. Respiratory difficulty may develop and even respiratory arrest occur. Prompt action under these circumstances may still be life-saving, if artificial respiration is instituted and immediate tapping of the abscess undertaken.

Brain abscess may be associated with congenital heart disease, particularly the Tetralogy of Fallot and other inter-ventricular defects, persistent truncus arteriosis and cor triloculare. The development of convulsions, focal neurological signs, and evidence of increased intracranial pressure in patients with congenital heart disease should arouse immediate suspicion of brain abscess. There may be no history of infection and there is usually no fever or clinical evidence of systemic inflammation. Headache, vomiting, lethargy and the progressive development of focal neurological sings, with or without mild meningismus, usually constitute the clinical picture. Surgical treatment combined with antibiotic therapy should yield a satisfactory number of successful results in such cases since the abscess is usually single.

The spinal fluid findings in cases of brain abscess usually consist of a mild pleocytosis with 50% or more of the cells being polymorphonuclear. Moderate elevation of the total protein is common; sugar is not usually altered and cultures are usually negative. The employment of a spinal puncture, when a brain abscess is suspected, must be regarded as hazardous as in the case of any intracranial mass and should be avoided if there are signs of increased intracranial pressure.

If brain abscess is suspected and there are sufficient localizing signs to proceed with reasonable certainty, the diagnosis may be established by tapping the abscess cavity through a bur hole. If the localization is not adequate, demonstration of the mass through arteriography or ventriculography may be necessary. Once the presence of the abscess is established and it is localized, treatment must be selected according to the problems presented by the case at hand.

Surgical Treatment

In the surgical treatment of brain abscess, tapping of the abscessed cavity repeatedly may be sufficient to effect a cure. This is the simplest form of treatment available and it is a general rule that the simplest

treatment is the best when it is effective. A small amount of radiopaque material may be introduced into the abscess cavity when the first tap is done, in order to follow by repeated x-rays the progress of decrease in size of the abscess. If sufficient scar to cause trouble remains from the residual capsule, this may be excised later.

A second method of attack is to introduce a catheter into the cavity after the initial tap. Constant drainage and the introduction of antibiotics is carried out through the catheter. With either tapping or catheter drainage, the abscess may tend to migrate toward the surface and become more accessible for future surgical extirpation.

Total excision of a brain abscess is clearly the ideal treatment in that it entirely eradicates the infectious focus from the cerebral tissue. Such extirpation was first found to be feasible in the case of old burned-out brain abscesses, often bacteriologically sterile, which were found accidentally when performing surgery for a suspected brain tumor. Now that heavy chemotherapy can be employed to prevent the spread of the infection, the excision of more acute encapsulated abscesses has become possible. While this is usually best carried out as a termination of treatment through tapping or catheter drainage, it may be occasionally the primary treatment of choice. Such excision is carried out as in any cerebral mass, choosing the shortest route to the encapsulated abscess through a silent area of the brain. By careful dissection of the softened brain around the gliotic capsule, the abscess may be delivered intact. The dura is then closed, the bone flap replaced and the scalp closed in the same way as in any craniotomy. Drainage is not employed unless there has been spillage of the abscess contents by rupture of the capsule in its delivery. Under these circumstances, it is probably best to leave a catheter in the cerebral defect and employ irrigation with antibiotic solutions in the same way that these are used in extradural and subdural abscesses. The brain tissue left behind is viable and has good blood supply, so that it is reached by chemotherapy employed parenterally as an adjunct to the surgical treatment.

Marsupialization, an older form of treatment, should not be forgotten. In this method, an encapsulated abscess near the surface, or more often one which had been encouraged to migrate to the surface, is opened and its edges approximated to the dura and scalp so that the capsule can turn itself inside out and extrude itself. This treatment may still be life-saving in very ill patients with large abscesses and a tough capsule, where tapping and drainage alone may not be adequate and excision would be too much for the patient to stand. Marsupialization used to be a very long process with a chronically draining wound persisting for weeks to months. With vigorous chemotherapy this can be greatly shortened by proceeding to excision as soon as the critical stage of the patient's condition has passed.

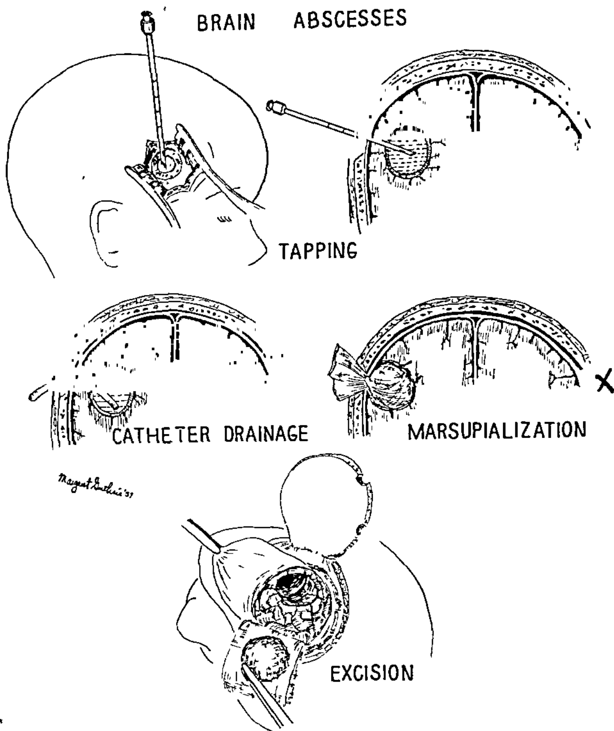


FIGURE 10. Methods of Surgical Management of Brain Abscess. Simple tapping with or without instillation of radiopaque material such as Thorotrast may be sufficient. After tapping, a catheter may be introduced into the abscessed cavity for more constant and prolonged drainage. Total excision of the abscess has been made possible by adjunctive chemotherapy and is the ideal treatment when feasible. Marsupialization may still be employed in patients with large abscesses with tough capsules when the patient is too toxic to permit primary excision.

Multiple abscesses may often result when the primary infection is in the lungs and hematogenous spread of the infection to the brain occurs. Under these circumstances there may be one or more abscesses large enough to give localizing signs and permit surgical drainage, but at autopsy many scattered small abscesses may be found, indicating that the prognosis was hopeless from the start. Any detectable abscess should nevertheless be drained in the hope that other infected emboli will be overcome by chemotherapy or local resistance. Occasionally multiple abscesses may occur, as complications of otitis and mastoiditis in both the temporal lobe and the cerebellum.

Tuberculous Spondylitis

Tuberculosis of the spine is most common between the ages of two and five years, and is a local manifestation of the disease reaching the vertebrae by hematogenous spread. It is characterized by destruction both of the intervertebral discs, especially in the thoracic area, and of the bone of the bodies of the vertebrae themselves. In a large percentage of cases, there are symptoms due to involvement of the spinal cord, usually beginning as weakness and stiffness in the legs. Paraplegia may result from compression of the cord due to the sharp knuckling of a kyphosis or from compression of the cord by epidural tuberculous tissue, in which case it may occur without deformity of the spine.

When there is tuberculous involvement of the cervical spine, the head is held very rigid. It may be extended or flexed or sometimes rotated or tilted to one side, and the child may support the head with the hands. Kyphosis is usually not marked in the upper cervical spine where the pain is referred to the occiput or neck, but with involvement of the lower segments the pain is referred to the arms or shoulders and marked kyphosis is not uncommon. Children with tuberculosis of the thoracic spine usually walk with great care to avoid jars. The body is commonly inclined forward and may be supported in this position by placing the hands on the knees. Sharp kyphoses appear in more rapidly progressive cases, whereas rounded curves are seen in less acute ones. Lumbar lordosis is often diminished when the tuberculous involvement is in the thoracic spine, but when it is in the lumbar spine itself, there is usually increased lordosis. Flexion of a hip due to psoas spasm because of accompanying psoas abscess may often be observed.

Roentgenological examination shows characteristic changes in tuberculosis of the spine. Destructive changes are first visible in the anterior part of the vertebral body, usually near the intervertebral disc. Later on collapse or wedging of one or more vertebral bodies occurs with reduction in thickness of the intervertebral discs. A soft tissue shadow around the area reflects reaction or abscess formation in the paraspinal structures.

It is to be hoped that the increasing success of tuberculosis detection techniques, and the use of streptomycin and isoniazid in the treatment of pulmonary tuberculosis will lessen the incidence of blood-born spread of the disease and, therefore, of spinal and spinal cord involvement. In the meantime, however, the chemotherapeutic agents may be utilized to make more radical surgical attack upon the bony abscesses and the spinal deformities possible without fearing local spread of the disease.

Surgical Treatment

Tuberculous abscess of the vertebral body (Pott's abscess) is best treated by curettage through a lateral paravertebral approach. In the thoracic area this may be readily accomplished by the resection of a rib or in the lumbar area along the psoas muscle. In this manner the abscess is approached anteriorly and the spinal canal not entered at all. The infected material is thoroughly curetted out and a gram of streptomycin powder put into the wound. Packing the cavity with bone chips at the time has been advocated by some, whereas others prefer fusion three or four months later after continuous administration of intensive anti-tuberculous medication.

In cases where the acute abscess has been eradicated or where the acute process has become arrested and a marked deformity is causing a chronic cord compression syndrome, judicious laminectomy in conjunction with spinal fusion or even resection of a protruding body to correct deformity and relieve cord compression may be occasionally undertaken.

When epidural tuberculous granulation tissue is causing compression of the spinal cord without accompanying deformity of the spine, laminectomy and curettage combined with chemotherapy may result in improvement in the neurological picture. There is tuberculosis of the laminae in most cases of this type, but there may not be any other actual tuberculous vertebral involvement.

Spinal Epidural Abscess

The spinal epidural space contains a number of blood vessels along with fat and loose connective tissue. The blood vessels provide an all-too-ready avenue for entry of blood-born infection, but the condition is nevertheless not a common one Furunculosis is most often the source of the infection, and the usual organisms are therefore staphylococci, but phlebitis, spinal osteomyelitis, pulmonary infections and decubitus ulcers are other origins. The infection spreads rapidly through the loose tissue of the epidural space and the cord may become surrounded by pus for half of its length or more. Although the infection does not extend into the leptomeninges, the spinal cord may show edema and softening.

The onset of spinal epidural abscess is characterized by intense pain in

SPINAL EPIDURAL ABSCESS

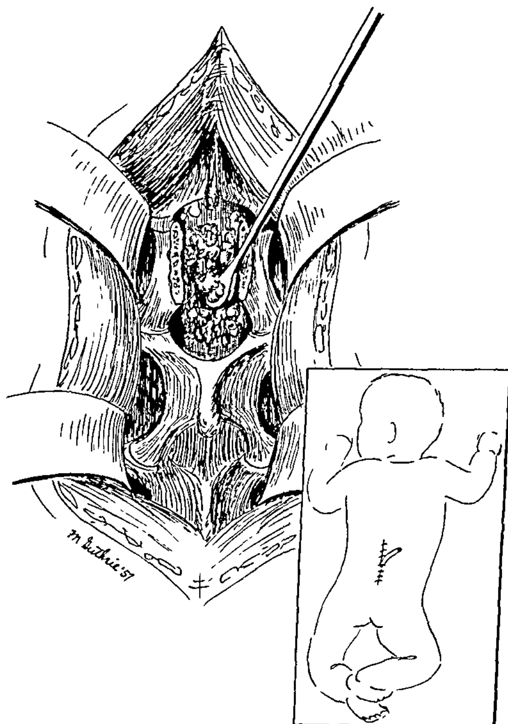


FIGURE 11. Acute Epidural Abscess—Surgical Management. This is a surgical emergency if a reasonable chance of recovery is to be afforded. Laminectomy must be sufficiently extensive to permit complete evacuation of pus and granulation tissue, and the abscess may have to be followed over a number of segments

the back radiating along the course of the spinal nerve roots. This is followed in a few days by rapidly progressive paralysis in the lower extremities. Fever, leukocytosis, rigidity of the spine, and local tenderness are generally present. This combination of signs and symptoms should immediately suggest the diagnosis. Spinal epidural abscess is a surgical emergency and only prompt laminectomy and drainage can result in restoration of neurological function. This should be supplemented by intensive chemotherapy.

A variety of infection in the spinal epidural space is extradural granuloma. While the organisms and origin are the same, the infection is not as virulent and there is usually no systemic evidence or local signs. The picture is rather that of gradually increasing weakness in the legs, similar to the clinical picture in a spinal cord tumor. Root pains will usually occur early. Laminectomy with excision of the granulomatous mass, which may have to be followed over a number of spinal segments, is the treatment of choice and should be supplemented by appropriate chemotherapy.

Abscess of the Spinal Cord

An acute myelitis developing in the course of septicemia, most commonly secondary to lung abscess, osteomyelitis, urinary tract infections, furunculosis and other infections due to staphylococci, may result in local softening with the affected segments of the spinal cord reduced to purulent material so that gray and white matter become indistinguishable. The clinical picture is that of a transverse myelitis with rapid onset, developing in association with septicemia or with a pyogenic infection in some part of the body. Intensive antibiotic therapy and surgical drainage may lead to recovery in rare instances, but the majority of recorded cases have had a fatal outcome.

Parasitic Cysts of the Central Nervous System

Infestation of the brain or the spinal meninges with the larval forms of the pork tapeworm (Taenia Solium) or the Taenia Echinococcus, an intestinal parasite, has become a neurosurgical problem of increasing importance. Cases are frequent in countries where hygiene is poor, but a significant number have also been reported from this country and England.

Cerebral Cysticercosis

Cysticercosis cerebri occurs in humans as a result of eating uncooked food which has become soiled with fecal material containing ova of the Taenia Solium. In the host's stomach the ovum develops into the embryo which works its way into the circulation and is carried to various organs, most commonly the eye, nervous system, or skin. The embryo then becomes

encysted and forms a cysticercus. There may be a solitary cyst in the brain, or multiple cysts may develop. Clinically, a single cyst gives rise to symptoms similar to brain tumor and may produce a focal seizure, hemiparesis, visual disturbance, cerebellar signs or may result in obstructive hydrocephalus when it grows within the ventricles. Multiple metastatic cysts result in diffuse cerebral swelling with elevated intracranial pressure, papilledema, visual loss, and mental changes resembling the syndrome of *increased intracranial pressure without tumor (pseudotumor cerebri)*. Finally, an intense reaction may occur in the meninges about the parasites with fibrous thickening giving the appearance of chronic meningitis, particularly in the basilar meninges, with production of hydrocephalus.

The diagnosis of cysticercosis must be kept in mind when the patient has been in a country such as Mexico, Peru or Chile where the disease is prevalent. Eosinophilia in the blood and spinal fluid may occur. A positive complement fixation test in the cerebrospinal fluid to an alcohol extract of porcine cysticerci may be helpful. Postive x-ray findings, due to multiple calcified cysts scattered through the brain may aid in the diagnosis.

In the recent years successful surgical attack has been carried out in cases of cysticercosis. Removal of a solitary cyst may be possible or shunt procedures may be feasible for the relief of hydrocephalus.

Cerebral Hydatid Cysts

Hydatid cysts of the brain occur more frequently in childhood than in adult life. Human infestation with the Taenia Echinococcus occurs only rarely in the United States, but is very common in Australia, New Zealand, Algiers, Tunis, Bulgaria, Romania, Iceland, Uruguay, and Argentina. The adult cestode is found in the intestines of the dog, wolf, or jackal. Many wild and domestic animals such as the sheep, ox, and pig as well as man may act as intermediate host. The chitinous shell of the ovum is digested in the stomach and the embryo liberated. It bores through the wall of the gut entering the portal vein and is carried to the liver and may spread by active migration or through the blood stream. Two per cent of all hydatid cysts are found in the brain. While usually solitary, multiple cysts may develop if there is rupture of a cyst attached to the wall of the left ventricle of the heart.

The appearance of cerebral hydatid cysts in children usually produces headache, vomiting, diminished visual acuity, hemiparesis, somnolence, and convulsive seizures. A characteristic feature may be unilateral bulging of the skull caused by pronounced thinning and flexibility of the bone. Papilledema is common. Symptoms suggesting intracranial tumor in a child from a country in which hydatid disease is prevalent should suggest

the diagnosis. Confirmatory evidence may be found in the presence of cysts in the liver or lungs, eosinophilia in the blood, or a positive Casoni cutaneous test. X-rays of the skull may show the characteristic changes in the cranial vault or show calcification of the cysts in rare instances. Angiography is safer than ventriculography when the diagnosis is suspected because it does not entail the risk of puncturing the cyst.

Neurosurgeons in countries where the disease is prevalent have obtained success in the removal of hydatid cysts in many instances. An ingenious technique has been devised where the cyst is forced to extrude itself by irrigating with saline between the cyst and brain substance. It is important to avoid rupture if contamination which may result in the recurrence of local infestation is to be prevented.

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CHAPTER XI

Intracranial Vascular Malformations

LESTER A. MOUNT

CHILDREN ARE subject to all the vascular anomalies or malformations found in adults, but which less frequently become symptomatic in the early years of life. For example, McDonald and Korb in reporting 545 cases of intracranial aneurysm found twenty-eight in children, and Walton in studying subarachnoid hemorrhage found eighteen under the age of twenty in a series of 320 cases.

The diagnosis of vascular anomalies and malformations in children is made by the same techniques used in adults, but variations in interpretation or methods do exist. Arteriosclerosis and hypertension are frequent causes of hemorrhage in adults but rarely in children. The presence of port wine stains or vascular nevi in the skin or vascular anomalies in the retina suggests the presence of a concomitant vascular anomaly in the brain. Dilation of arteries and/or veins of the scalp or face, which represent arterial, venous, or arteriovenous anomalies, suggests the possibility of the same anomaly in the brain. Dilatation of the veins of the scalp also suggest an intracranial arteriovenous communication, especially if they pulsate. Enlarged veins of the scalp may be the result of superior sagittal sinus thrombosis or an enlarged parietal or mastoid emissary. A pulsating exophthalmos hints of the probability of an arteriovenous fistula between the internal carotid artery and cavernous sinus. Dilatation of the carotid arteries on one side may indicate the presence of an arteriovenous communication intracranially, whereas absence of pulsation intimates thrombosis or absence of the carotid arteries. A soft mass along the midline of the scalp which becomes prominent on bending over or straining at stool is indicative of sinus pericranii.

Cranial bruits may be heard in as many as 10 to 15% of children without intracranial lesions according to some authors and an even higher per cent in children under the age of five. In adults they are always of pathological significance. Cranial bruits are most frequently heard in patients who have arteriovenous angioma or fistula and less often in patients who have arteriovenous angioma or fistula and less often in patients having saccular

aneurysms, brain tumors, tumors pressing upon the major arteries in the neck, coarctation of the aorta, or even a highly vascularized thyroid gland.

X-rays of the skull may show enlarged vascular channels or intracranial calcifications. The electroencephalogram is of little value other than showing focal abnormalities in some instances. The visual fields will show defects if the pathways or visual cortex is involved.

Arteriography is the main diagnostic test and the technique is described under the chapter entitled diagnostic techniques. Venography is at times necessary in order to make a diagnosis of a vascular malformation and its technique is also presented under diagnostic techniques.

Vascular Anomalies of the Scalp

Vascular anomalies of the scalp are important to the neurosurgeon primarily because they frequently reflect concurrent anomalies within the brain.

These anomalies, as well as those intracranially, are apt to be multiple in type as well as location. Some, such as the arteriovenous malformations, may involve the scalp, the skull, the meninges, and the brain. Large arteries in the scalp with a vigorous pulsation may indicate only that they are assisting in supplying the brain or an intracranial arteriovenous anomaly with blood by means of these collateral vessels in the scalp. Large distended veins in the scalp may represent accessory channels of drainage of blood from a large intracranial arteriovenous angioma, or of drainage of blood from the brain when there is thrombosis of the superior sagittal sinus.

The vascular anomalies of the scalp are arterial, venous, capillary, or arteriovenous. These require no discussion in a text of neurosurgery, but two varieties of venous anomalies warrant further description. These are sinus pericranii and persistent large emissary veins.

Sinus Pericranii

Sinus pericranii is a venous sinus beneath the pericranium which is connected with one of the dural venous sinuses, usually the superior sagittal sinus, by small openings through the skull. It is not visible or palpable unless the intracranial pressure is raised as in crying or straining at stool, and it also appears on bending over. X-rays, as in Figure 1 show a soft tissue mass and an emissary vein opening through the skull. It can readily be demonstrated by injecting hypaque into it as in Figure 2, but this is usually unnecessary. It is treated surgically by removing the endothelial sac and packing the communications through the bone with bone wax.

Case 1. A Patient With Sinus Pericranii Accompanied by Other Vascular Abnormalities

A four and one-half year old child was admitted because of the presence of a soft tissue mass in the right forehead since birth, a spastic paresis of the left arm and leg since birth, and two episodes of nausea, vomiting and occipital pain eight and five days before admission. Immediately after birth the mother noted a pink birthmark about 4 cm. long and 1 cm. wide just to the right of the midline of the forehead. The bony defect could be felt beneath this. It was also noted that this area pulsated. It seemed to remain the same size for three and one-half years and then the skin became darker in color. Eight months before the child was seen, it began



FIGURE 1 Radiolucent area in the outer table of the frontal bone beneath the sinus pericranii (A defect need not be associated with sinus pericranii.)

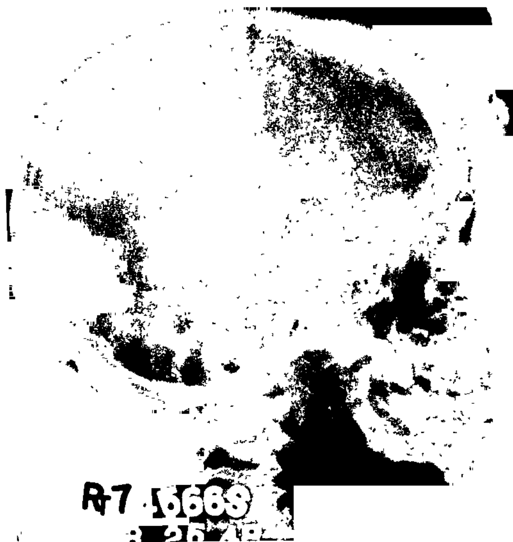


FIGURE 2. A venogram of a patient with sinus pericranii performed by direct injection of diodrast into the anomaly. The needle and syringe are also seen. The portion of the sinus immediately adjacent to the bone was not seen in the venogram.

to grow. It was noted to be more prominent on sneezing, coughing, or laughing and was not painful to touch.

Spastic paresis of the left arm and leg, which had been noted at birth, had shown some improvement in the three years before the patient was examined and during which time the patient had had exercises and massages. There were no other important points in his history except for the presence of a hairy dermoid cyst which had been removed from the outer canthus of the right eye at the age of three months.

Examination showed a fairly well developed child who had telangiectatic vessels over both cheeks and a bulging, tense, pulsating, soft tissue mass slightly pigmented, measuring 7 x 3 x 2 cm. over the right forehead. It was compressible but did not transilluminate. There is a palpable defect measuring 5 x 1 cm. in the bone underlying this mass. A loud bruit

could be heard over the entire cranium, loudest above the right ear. There was a spastic hemiparesis involving the left upper extremity to a greater extent than the lower extremity. The deep tendon reflexes were hyperactive and the Babinski and Chaddock signs were positive on the left.

X-rays of the skull revealed a scalp tumor which communicated through the midfrontal bone with the cranial cavity, suggestive of a lesion of the sinus pericranii type, but also a more extensive vascular malformation or angioma.

Venogram films were made after the injection of diodrast into the scalp tumor in the frontal region. The first film was made near the end of the injection, while the second film was made approximately ten seconds later. The contrast material outlined a markedly dilated tortuous vascular channel which had an appearance suggesting a large varicosity. In addition, there was stasis of the contrast material in the varicosity ten seconds after the injection. An efferent vein which leads downward in the soft tissues of the forehead toward the face was seen on both films.

Carotid arteriography showed a large arteriovenous angioma involving the entire right hemisphere.

The vascular mass of the scalp was treated by turning a flap of the scalp around the area. When the flap was reflected forward two large thin-walled veins were seen passing through the bone to the mass. These were ligated and as much as possible of the thin-walled vascular mass was removed. The vascular connections with it were ligated so that the part remaining was in a collapsed state. The remaining portion could not have been removed without also removing the overlying scalp.

The child was readmitted one and one-half years later because of an increase in the frequency of his convulsive seizures, and coma of twenty-four hours duration following an epileptic attack.

The significant findings on examination were a semiconscious child with a temperature of 104° F, pulse 60, and respirations 40. He responded to painful stimuli and only blinked his eyes to auditory stimuli. There was underdevelopment of the left side of the trunk and left extremities. A loud bruit was heard over the entire cranium, but was best heard over the right parietal region. The noise could be eliminated by compression of the carotid vessels on the right side of the neck. The left extremities were spastic. The deep tendon reflexes were hyperactive on the left side, with a bilaterally positive Babinski sign. The nasal margins of the optic discs were indistinct. The left pupil was larger than the right, but both reacted well to light, the right better than the left.

Arteriography showed an increase in the size of the arteriovenous malformation. The patient was treated by ligation of common and external carotid arteries. This greatly reduced the intensity of the bruit but the patient did not improve.

Comment. This case illustrates a combination of vascular malformations. On inspection, this patient had telangiectasis of the cheeks, a sinus pericranii, and a bruit could be heard over the head. In addition, the arterio-

gram revealed an arteriovenous malformation involving the entire right cerebral hemisphere.

Persistent Large Emissary Veins

Persistent large emissary veins, as the name indicates, are large communications between the intracranial and extracranial venous systems usually by way of the parietal or mastoid emissary veins. As a result of the large communication, when the patient raises his intracranial pressure by crying, straining at stool, lifting, or upon bending over, the veins of the scalp become distended like a caput medusae. Venography can readily be done, but with a history of such a condition since birth, this is unnecessary unless blockage of the intracranial venous sinuses is suspected. Enlarged veins of the scalp secondary to an intracranial arteriovenous angioma must also be considered. Arteriography is necessary in such cases. The treatment is double ligation and section of the communication at the point where the veins enter the bone.

Intracranial Vascular Diseases

The intracranial vascular diseases of surgical interest, excluding the vascular tumors, are angiomas and aneurysms. The angiomas are arterial, venous, capillary and arteriovenous. Other descriptive names are also used, such as racemose, cavernous, varix, varicosity, etc. They may be further classified according to location as meningeal or parenchymatous. The aneurysms are usually saccular in type and most often arise from the major arteries at the base of the brain, usually from the internal carotid artery or, the circle of Willis, the middle cerebral artery, the vertebral basilar system, and rarely from vessels located in the substance of the brain.

Spontaneous subarachnoid hemorrhage and spontaneous intracerebral hemorrhage, while not disease entities, are important conditions to the neurosurgeon and will be discussed as special topics.

The Intracranial Angiomas

The angiomas are really malformations and not tumors as the name suggests. They grow, due to enlargement of the involved vessels or to the implication of additional vessels. Although they are formed early in the developmental period only 35% become symptomatic under the age of twenty. The three most common first clinical manifestations are hemorrhage, epilepsy, and periodic headaches. Other symptoms and signs are dependent upon the location of the lesion in the brain. In the larger angiomas, especially the arteriovenous ones, a bruit is often heard by the patient and the examiner.

The x-ray shows calcification within the lesion in one-fourth of the

patients. Arteriography is necessary to delineate the size, shape, and position of the pathological process, to study the collateral circulation, and to determine the presence of multiple lesions. Bilateral carotid arteriography is necessary in all cases. Vertebral arteriography is necessary when carotid arteriography is negative or when a lesion is suspected in the distribution of the vertebral-basilar system.

The treatment is surgical removal when it can be accomplished without a serious neurological defect. X-ray therapy is of little value. When complete removal is impossible, strategic clipping of as many of the feeding arteries as possible may be helpful in destroying part of the lesion, in reducing the possibility of hemorrhage and in alleviating some of the symptoms, especially the headache.

The surgical technique of handling these cases is easy since good arteriograms, stereoscopic if necessary, will show the feeding arteries. When these are ligated the lesion will collapse like a deflated balloon. On the other hand, if the lesion is entered before the feeding vessels are clipped the bleeding can be profuse.

Carotid ligation has been performed in patients having arteriovenous angiomas when the intracranial approach is not feasible. This is a palliative procedure which may temporarily relieve some of the symptoms. Arteries can be ligated in stages and even bilateral common internal and external carotid ligations have been performed for this condition.

Arterial angioma consists of a conglomeration of arteries of varying sizes which are frequently thin-walled. They are found on the surface and may extend deeply into the brain. Frequently, they are associated with a capillary angioma and/or with a concomitant venous angioma with direct communications, thereby forming an arteriovenous angioma. Atrophy or lack of development of the brain in the region of the angioma may occur. Symptoms and signs are the result of these changes in the brain; or they may be due to rupture of the angioma and hemorrhage into the substance of the brain, and/or into the subarachnoid or subdural spaces. Frequently no symptoms or signs are present until hemorrhage occurs. The clinical features are dependent upon the area of the brain involved and there may be convulsions, mental disturbances, hemiparesis, incoordination, reflex abnormalities, sensory changes, aphasia, or visual disturbances. X-rays of the skull are usually normal. The visual fields or electroencephalogram may show a focal lesion but arteriography (Figure 3) is necessary to make a positive diagnosis.

The treatment is surgical removal whenever this can be accomplished without leaving a serious neurological defect. Strategic clipping of some of the arteries supplying the angioma may be helpful when total removal is inadvisable because of location.

Case 2. A Patient Having an Arterial Angioma in the Parieto-Occipital Region

This 11 month old child was admitted because of a right hemiparesis and right-sided seizures. The gestation period and delivery were normal. Nothing unusual was observed until the age of two months when she began to have right-sided tonic and clonic seizures starting in the fingers, then the face and lower extremity, with loss of consciousness, and interestingly enough, deviation of the head and eyes to the left side. Until anticonvulsive medication was started, the seizures lasted from one to three



FIGURE 3. An arteriogram and a pneumoencephalogram of a patient having an arterial angioma and cerebral atrophy or hypoplasia. (a) The arrows point to the arterial angioma which is supplied by the posterior cerebral and posterior parietal arteries. There is a suggestion of a capillary component. The abnormal shadows in the frontal region are artifacts.

hours. The child's developmental history and psychometric studies revealed retardation.

On examination there was a large port wine stain over the left upper eyelid, left forehead, and left side of the scalp extending back to the occipital region. The child was able to move all of her *extremities*, but



FIGURE 3. (b) In the anterio-posterior arteriogram in the late arterial phase, the vertical arrow points to angioma and horizontal arrow indicates that the anterior cerebral arteries are shifted to the side of the angioma. The pneumoencephalogram films Figure 3 (d) and (e) show that this is due to atrophy

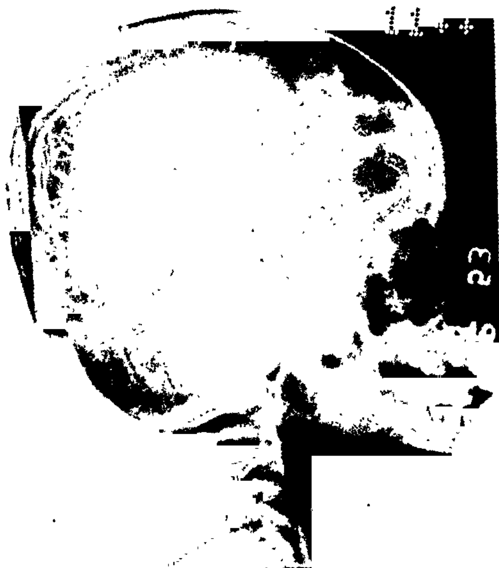


FIGURE 3. (c) The arrow points to the superior sagittal sinus which is present only in the posterior one-third of the skull.

the right much less than the left. The tone was increased in the right extremities, more so in the upper extremities than in the lower. Coordination was poor in the right upper extremity. There was slight hyperreflexia in the right upper extremity. Painful stimuli evoked less of a response on the right than on the left. The cranial nerves were normal. Palpation and auscultation of the skull was normal.

X-rays showed the skull to measure 15.6 cm. in length and 14.6 mm. in width, indicating a brachycephalic type of skull with a rather flat occipital region. There was no evidence of widening nor premature closure of the sutures. A large wormian bone was seen in the lambdoidal suture, which is a normal variant. The sella turcica, the orbit, and the petrous pyramids were normal. No abnormal intracranial calcifications were seen.

The spinal fluid protein was 50 mg. %.

The arteriogram revealed good filling of the internal carotid and its intracranial branches. The posterior cerebral artery filled from the carotid

system and was larger than average, being about the size of the trunk of the middle cerebral artery. In the occipital region, there were combinations of small arteries and large capillaries, an angiomatous type of malformation without the usual large caliber vascular channels.

An unusual finding was failure of the vessels to reach the superior margin of the brain as would be expected normally, being separated from a distance of approximately 2 cm. from the inner table of the skull along the midline. For this reason the presence of a collection of fluid or possibly a subdural hematoma on the superior surface of the brain could not be excluded. Cerebral atrophy with external hydrocephalus could produce the same picture. Another finding of great interest was the superior longitudinal sinus filling only in its posterior one-third. No contrast material was seen in the anterior two-thirds. Evidently, most of the circu-



FIGURE 3 (d) Lateral pneumoencephalogram films reveal marked atrophy or lack of development, especially in the parietal and occipital regions on the left side

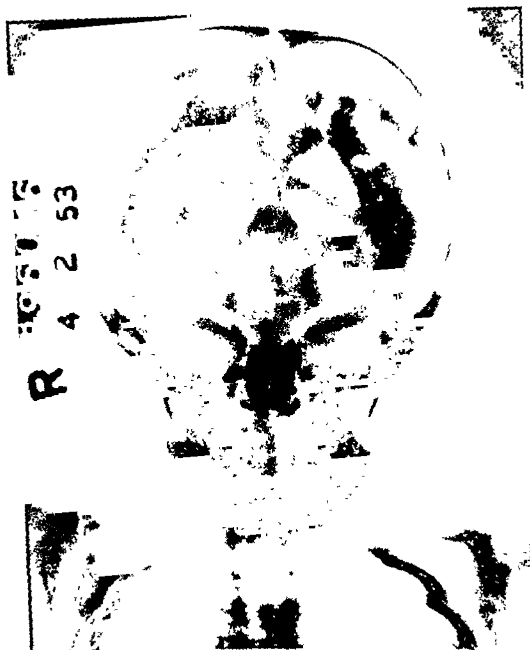


FIGURE 3 (c) Posterior-anterior pneumoencephalogram films reveal marked atrophy or lack of development, especially in the parietal and occipital regions on the left side.

lation in the venous phase was through the anastomatic veins of Labbe and down the lateral sinus without filling of the transverse portion. One would suspect there may be agenesis or thrombosis of the anterior portion of the longitudinal sinus. Furthermore, there were no superior cerebral veins extending upward in the frontal and anterior parietal regions.

The pneumoencephalogram made after the replacement of 60 cc. of cerebrospinal fluid by oxygen showed that both lateral ventricles were slightly larger than average, the right being larger than the left. The enlargement was symmetrical The third ventricle was in the midline and was abnormal in shape, the aqueduct and fourth ventricle were normal. A large collection of gas was seen just above and behind the aqueduct

and third ventricle. This represented a huge cisterna ambiens rather than a sixth ventricle. The suprasellar cisterns were slightly wider than average, and the lateral arms of the cisterna ambiens were likewise more prominent than usual. This was noticed more on the left side than on the right. Huge subarachnoid sulci were present in the parietal region and in the posterior half of the frontal lobe. There was actually a space filled with air between the inner table of the skull and the surface of the brain which measures 1.5 cm. This would obviously account for the wide separation between the inner table of the skull and the cerebral vessels as seen in the arteriogram.

Psychometric examination showed a little retardation.

Summary Diagnosis: (1) Arterial angioma left parieto-occipital region.

(2) Considerable atrophy of the brain, primarily of the left cerebral hemisphere.

(3) Mental deficiency.

(4) Agenesis of anterior two-thirds of superior sagittal sinus.

Venous angiomas are congenital malformations of a group of thin-walled vessels of various sizes, types and locations. They are found in the meninges, including the dura as well as the leptomeninges and may extend deeply into the brain. They may be simple varices found only on the surface of the brain, or large cavernous or racemose malformations extending deeply into the brain as far as the ventricle or basal ganglia, and may be continuous with a venous angioma of the scalp. Cerebral tissue is found among the intracerebral masses of the veins.

The symptoms and signs, when present, are dependent upon the location of the lesion. The most common finding is focal epilepsy followed closely by mental deficiency and hemiparesis. Hemiplegia may follow a convulsive seizure and is due to thrombosis within the lesion, though occasionally it may be due to hemorrhage. Sensory changes, visual field defects, or aphasia may be present. There is no evidence of increased intracranial pressure unless in rare instances the malformation causes blockage of the ventricular system with resultant hydrocephalus.

The skull roentgenograms show no abnormality except that occasionally, curvilinear streaks of calcium are found in the lesion. The electroencephalogram may or may not show any abnormality. However, even if it did reveal a focal abnormality it is not helpful in determining the type of pathological process. The serialographic arteriogram clearly demonstrates the size, shape, position and type of pathological process which is present as in Figure 4 (a) through (d).

The only treatment is surgical excision of the lesion if it is located in an area in which removal will not result in a serious neurological defect in the patient X-ray therapy is [illegible] little or no valu[illegible] [illegible]se, the treatment is purely symptomatic [illegible] [illegible]ulsant [illegible] seizures).



FIGURE 4. These are arteriographic films of a patient with venous angioma, but there is also an associated capillary component. (a) The lateral serialographic anteriogram showing an enlarged precunial branch of the anterior cerebral artery and a smaller post-rolandic branch of the middle cerebral artery supplying blood to a capillary angioma.

Case 3. Venous Angioma

A nine and one-half year old child was found lying on the floor unable to move his right extremities and had difficulty in speaking.

Examination showed a semi-conscious child barely responsive to painful stimuli. There was right flaccid hemiplegia with a right Babinski and a right central facial paresis. X-rays of the skull were normal. An electroencephalogram revealed a left frontal, temporal, parietal, and occipital focus. The lumbar spinal fluid was under an initial pressure of 390 mm. of water with 27,000 cremated red cells.

Bilateral carotid arteriography twenty-six days after his hemorrhage, revealed on the left side a vascular stain beginning to appear in the frontal region at one and one-half seconds and increasing in intensity and persist-

A ten lead electroencephalogram with monopolar and bipolar recording revealed a definite right temporal focus of one to two second high voltage activity. It was basically a disorganized record.

An arteriogram as in Figure 5 (a) and (b) showed cross filling of the left anterior and middle cerebral vessels from the right carotid artery through the anterior communicating artery. The right vertebral, basilar, and posterior cerebral arteries also were filled. The right middle cerebral artery was well filled with contrast material only to a point about 1½ cm. from the terminal bifurcation of the internal carotid artery, and beyond this point there was only some faint filling of the vessels. This finding could be caused by a thrombosis of the right middle cerebral artery. In addition, the anterior cerebral vessels were displaced toward the right,



FIGURE 5. Sturge-Weber Syndrome. (a) A lateral arteriogram showing a telangiectatic area in the parietal region as indicated by the arrow. It also extends in the frontal region.



FIGURE 5. (b) The anterio-posterior view reveals the shift of the anterior cerebral arteries to the right side as indicated by the horizontal arrow and the partial thrombosis of the middle cerebral artery as indicated by the other arrow.

which indicated loss of tissue of the right hemisphere due to atrophy or hypoplasia. In the arterial and venous phases some dilated, irregular and tortuous vessels were seen in the upper portion of the right parietal region, and to a lesser extent in the frontal region, which indicated the presence of an angioma of the telangiectatic type. The inferior longitudinal sinus was quite long and extended far anteriorly. The straight sinus appeared quite narrow, which may be an indication that the venous drainage was in an anterior direction instead of the usual posterior downward direction.

In summary, the examination showed evidence of an angioma of the telangiectatic type in the upper right parietal region and possible thrombosis of the right middle cerebral vessels, as well as hypoplasia or atrophy of the right cerebral hemisphere as evidenced by deviation of the anterior cerebral vessels toward the right.

A 65 cc. pneumoencephalogram as shown in Figure 5 (c) revealed the

lateral ventricles to be moderately dilated with the right ventricle slightly larger than the left and somewhat higher in position. The ventricles were shifted toward the right approximately 8 to 10 mm. beyond the midline. It was noted that the patient had a sixth ventricle, a persistent cavum vergae, which is a congenital anomaly of no clinical significance. There was evidence of marked cerebral atrophy on the right, particularly in the anterior and posterior regions with marked widening of the sulci. Some of the basal cisterns were also unusually wide. Air was seen in the subdural space.

The total protein in the cerebrospinal fluid was 216 mg. %.

The child was placed on anticonvulsive medication.

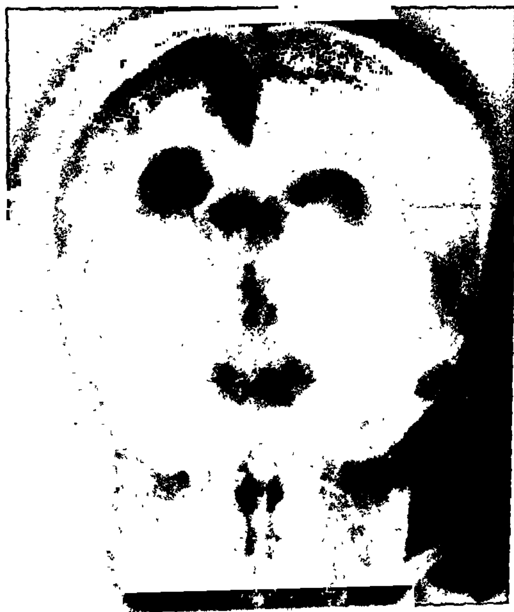


FIGURE 5. (c) The pneumoencephalographic films demonstrate the atrophy of the brain as evidenced by the enlargement of the ventricle and the widening of the sulci.

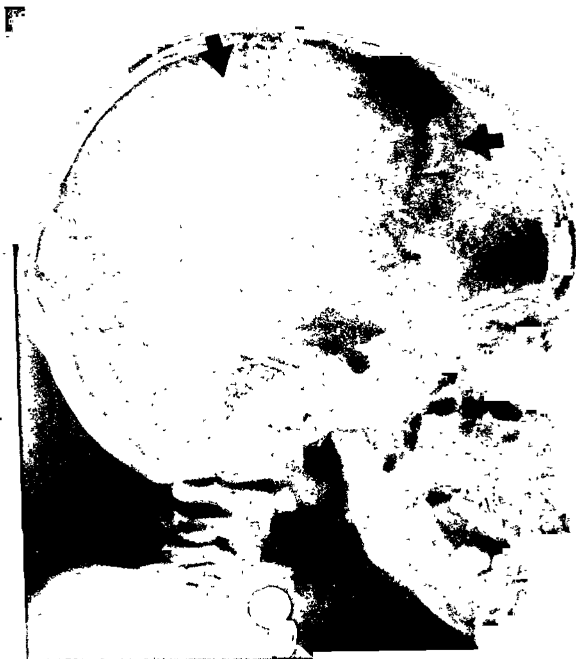


FIGURE 5. (d) The plain x-ray film taken two years after the previous films show the calcification in the cortex of the brain in the frontal and parietal regions.

At the age of thirty-two months he was able to walk with a little assistance. The patient was able to sit and stand by himself and able to speak a few isolated words. He was not toilet trained and was definitely mentally retarded.

There was a spastic left hemiparesis with left hyperreflexia and a left positive Babinski sign. An X-ray of the skull at this time showed extensive calcification of the cortex in the frontal and parietal regions.

The final diagnosis was:

(1) Sturge-Weber syndrome

(2) Thrombosis right middle cerebral artery, and
(3) Severe cortical atrophy, right greater than the left.

The Intracranial Arteriovenous Communications

The intracranial arteriovenous communications are of two types: (1) Arteriovenous fistula which are most frequently traumatic and are almost always within the cavernous sinus, and (2) Arteriovenous angioma or varix which is congenital and may be found in any part of the brain but most frequently involves the middle cerebral artery.

Arteriovenous fistula between the internal carotid artery and the cavernous sinus is the result of trauma with a fracture of the base of the skull and tearing of the internal carotid artery within the cavernous sinus, or the result of rupture of an aneurysm of the internal carotid artery within the cavernous sinus. It is characterized by the sudden onset of a noise in the head, synchronous with the pulse, followed by increasing prominence of one and occasionally both eyes, diplopia, and later impairment of vision and pain in the eye and forehead. A bruit is heard over the closed eye. The symptoms and signs are caused by the direct pouring of arterial blood into the cavernous sinus with distension of the cavernous sinus, as well as the veins which communicate with this sinus, especially the ophthalmic veins. It is the distention and pulsation of the ophthalmic veins which cause the pulsating exophthalmos. The pressure on the ophthalmic branch of the trigeminal nerve, the oculomotor nerve, the trochlear nerve, and the abducens nerve as they pass through the cavernous sinus orbit causes the reduced corneal reflex, dilated pupil, and extraocular muscle weaknesses. If there are wide communications through the circular sinus with the opposite side of the cavernous sinus, the symptoms and signs will be bilateral. If there is no communication between the cavernous sinus and the ophthalmic veins on the side of the fistula, the symptoms and signs will all be on the opposite side.

The bruit usually can be greatly reduced by compression of the carotid artery in the neck on the same side and abolished by compression of both carotids. If it cannot be abolished by bilateral carotid compression, a rich supply must be contributed through the posterior communicating artery from the vertebral basilar system.

The differential diagnosis rests among congenital arteriovenous angioma within the orbit, aneurysm of the internal carotid artery, aneurysm of the ophthalmic artery, defect in the roof or posterior wall of the orbit, tumor of the orbit and cavernous sinus thrombosis.

The diagnosis of carotid cavernous sinus fistula is made by carotid arteriography, in which the films are characteristic. In the arterial phase the cavernous sinus is well demonstrated as are the large ophthalmic veins, the internal jugular vein, and often the middle cerebral vein.

Treatment is ligation of the internal carotid artery in the neck and intracranially proximal to the posterior communicating artery. Ligation of the ophthalmic artery at the same time is desirable. Occasionally, ligation of the carotid artery in the neck only is sufficient, but this cannot be depended upon. The internal carotid artery is ligated in the neck with a Selverstone clamp so that the artery can be opened should the patient show signs of insufficient collateral circulation. Fortunately, this is necessary infrequently. The intracranial ligations should be done within a week of the cervical carotid ligation.

Dandy has advocated ligation of the internal carotid artery intracranially and then in the neck of the same operation. The rationale is that the intracranial ligation prevents backflow of the collateral circulation via circle of Willis into the fistula. This is a safe procedure if the arteriogram on the side of the fistula shows all the contrast material going into the fistula.

In the absence of therapy all the symptoms become progressively worse and blindness ultimately occurs.

The arteriovenous angioma or varix is composed of one or more enlarged arteries supplying a rich amount of blood to a network of varying sized and shaped thin-walled vessels emptying directly into veins which are dilated with completely arterial blood or swirls of mixed arterial and venous blood. They are usually located supratentorially but are also found in the posterior fossa. They may be superficially placed or extend in a wedge shaped manner toward the ventricle. They are supplied by one or more major arteries of the brain and frequently by two adjacent arteries, for example, the middle and the anterior cerebral arteries. Occasionally in midline lesions the anterior, middle, and posterior cerebral arteries on both sides will supply blood to the arteriovenous communication.

The symptoms and signs are varied and again are dependent upon the locations of the pathological process. The frontal lobe, being the largest lobe, is most frequently the location. Hence, focal convulsive seizures are the most frequent early symptom. There may be weakness and smallness in the extremities on the opposite side. Mental changes or aphasia are sometimes present. Sensory seizures, numbness, poor coordination, and impaired sensation are found in varying combinations when the parietal lobe is involved. Visual field defects are present when the occipital or temporal lobes are involved. Temporal lobe fits may be present and, aphasia in addition when the lesion is in the dominant temporal lobe. Ataxia, gait disturbances, and nystagmus are found when in the posterior fossa. The patient frequently complains of a noise in his head, and a bruit may be heard over the entire head, or over closed eyes, or in the neck over the carotid arteries. The bruit is reduced or abolished by compression of the carotid in the neck on the involved side when the lesion is supratentorial

and is usually abolished by bilateral carotid compression. If the bruit is unaffected by carotid compression or is made louder, the lesion must be in the distribution of the vertebral-basilar system. Greater pulsation may be seen and palpated in the common carotid artery on the involved side. Dilatation of the veins of the scalp is sometimes seen. This may not be so apparent until the hair is shaved. Vascular lesions in the skin or retina are occasionally found. The veins of the retina may be enlarged. Increased intracranial pressure is often found with resultant headache, nausea, vomiting, visual disturbances, and elevated spinal fluid pressure. Frequently these lesions rupture, causing intracerebral, subarachnoid, or subdural hemorrhage and, at times, death. Thrombosis in the lesion occasionally occurs. Hydrocephalus is sometimes present if the lesion is in the midline and obstructs the flow of the cerebrospinal fluid.

Roentgenograms of the skull may show enlarged diploic channels or calcium in the wall of some of the vessels in the lesion. The electroencephalogram may demonstrate a focal abnormality, but arteriography is necessary to delineate the pathological process in order to determine the therapy.

Treatment is resection of the lesion whenever possible. First, the arterial supply to the angioma is ligated and the vessels collapse like a punctured tire. If this does not happen there must be additional arteries contributing to it. Induced hypotension or hypothermia is helpful but is not necessary when surgical therapy is performed.

Ligation of the carotid or vertebral arteries in the neck may temporarily delay the progress of the disease as in Case 1. However, this should not be done unless total excision of the lesion would increase the neurosurgical deficit. Both the external and internal carotid arteries on both sides have been ligated, in stages, for such a lesion.

What will happen if no surgical therapy is performed? It is well recognized that the danger of a fatal hemorrhage from an angioma is much less than that of an intracranial aneurysm, and patients have had repeated hemorrhages without permanent neurological defects. However, there is always danger of hemorrhage with increase in the neurological deficit, or death. It is, therefore, the best judgment to remove the pathological process whenever there is a reasonable chance of success.

Case 5. Bilateral Arteriovenous Angioma Involving the Anterior Middle and Posterior Cerebral Arteries and Secondary Hydrocephalus

An 11 month old baby was noted by her parents to have some engorgement of the vessels on the right side of the head and some asymmetry of of the head at the age of 5 months One week before admission the parents noted that the child could not move her left extremities, and 9 hours later

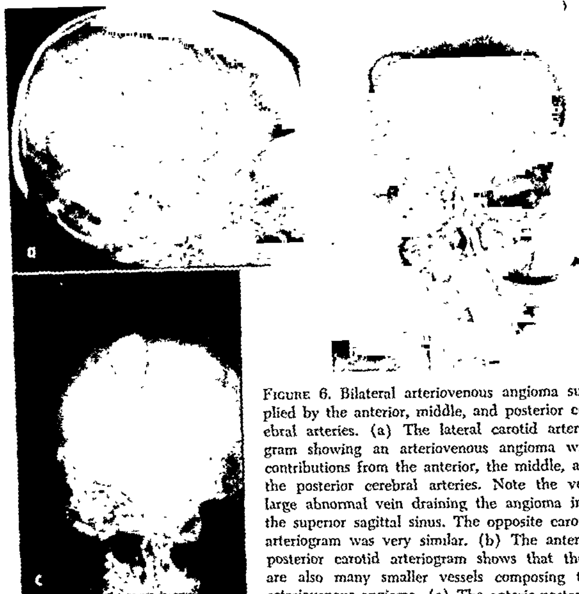


FIGURE 6. Bilateral arteriovenous angioma supplied by the anterior, middle, and posterior cerebral arteries. (a) The lateral carotid arteriogram showing an arteriovenous angioma with contributions from the anterior, the middle, and the posterior cerebral arteries. Note the very large abnormal vein draining the angioma into the superior sagittal sinus. The opposite carotid arteriogram was very similar. (b) The anterio-posterior carotid arteriogram shows that there are also many smaller vessels composing the arteriovenous angioma. (c) The anterio-posterior vertebral arteriogram demonstrates in one film the bilateral location of the angioma and the very large basilar artery which is sending branches to it.

she had a left-sided tonic and clonic seizure lasting 45 minutes. She had only one other seizure which occurred on the same day. Shortly after this attack the child began to move the left lower extremity.

Examination revealed a child with an enlarged head who was able to walk with some help. The anterior fontanelle was full. The deep tendon reflexes were hypoactive but equal in the upper extremities and were slightly greater in the right lower extremity than in the left. There was a positive Babinski on the left side. Sensation was normal. No abnormalities were observed in the cranial nerves. A loud bruit was heard over the entire head X-rays of the skull showed the head to be slightly enlarged. An electroencephalogram showed no abnormality. A right carotid arteriogram revealed a large arteriovenous angioma between the right

middle and anterior cerebral arteries which drained into an abnormal vein which extended upward from the region of the incisura to the superior sagittal sinus. The left carotid arteriogram showed a similar arteriovenous angioma with contribution by way of the anterior middle and posterior cerebral arteries. There was elevation of the anterior cerebral artery suggesting hydrocephalus. The vertebral arteriogram showed both posterior cerebral arteries to be contributing a rich supply to the angioma.

The cause of the hydrocephalus was believed to be obstruction of the aqueduct of Sylvius by the large arteriovenous mass. See Case I also.

Intracranial Arterial Aneurysm

Intracranial arterial aneurysms are weakened areas in the walls of arteries of the brain which have bulged out like a blister.

The etiology in children is a deficiency in the internal elastic membrane or the muscularis, infarction or infection in the arterial wall secondary to sterile or infected emboli (usually from the heart), or rarely arteriosclerosis, even in children. A greatly increased arterial pressure from coarctation of the aorta is a contributory cause especially when a weakness in the wall is already present. Intracranial aneurysms are often associated with polycystic kidneys and occasionally with coarctation of the aorta. They are most frequently found on the circle of Willis, on the internal carotid artery.

The symptoms and signs produced by an aneurysm are due to local pressure on the adjacent structures. When rupture occurs hemorrhage into the subarachnoid and/or subdural space, or into the brain, causes local and general increased intracranial pressure and also arteriospasm. The onset is sudden in 90% of cases. Anything which causes the elevation of the blood pressure temporarily may be a precipitating cause. This includes exercise, straining at stool, excitement, anger, etc.

Since aneurysms most frequently arise from the internal carotid artery, the adjacent structures which may be encroached upon are the third cranial nerve, the fourth, the sixth, the ophthalmic branch of the fifth and sometimes the optic nerve or chiasm. As a result of such compression, the patient will complain of pain in the eye and forehead and double vision, and on examination will have ptosis, dilatation of the pupil, absence of reaction of the pupil to light, extraocular muscle weaknesses, reduced corneal reflex, and impairment of vision. Various combination of these clinical findings may be present. The aneurysms within the cavernous sinus are likely to cause all of these symptoms and signs while those outside the cavernous sinus usually cause only pain in the eye and forehead and third nerve signs unless they are very large. Aneurysms in other locations are not so likely to cause symptoms by local pressure, but hydrocephalus from obstruction of the aqueduct of Sylvius has been reported.

Often hemorrhage is the first signs of the presence of an intracranial

aneurysm. When an aneurysm ruptures into the subarachnoid space, the patient has the sudden onset of headache, nausea, vomiting, dizziness, and frequently may collapse, progressing to coma. Upon recovery of consciousness, the patient had headache, vomiting, dizziness, photophobia, confusion, stiff neck and a positive Kernig's sign. Sweating and pain in the back and lower extremities are sometimes present. The pulse rate is usually elevated as is blood pressure and the temperature soon becomes elevated. Subhyaloid hemorrhages and papilledema often occur later. Frequently there are no symptoms and signs to indicate the location of the lesion. The resultant clot may exert pressure upon the adjacent structures. This is particularly true when an aneurysm of the internal carotid artery ruptures. The clot can then press upon the 2nd, 3rd, 4th, 5th and 6th cranial nerves as described in the preceding paragraph. Ruptured aneurysms of the middle cerebral artery may produce focal or generalized convulsive seizures, hemiparesis, hemihyperreflexia, unilateral sensory changes, aphasia, apraxia, and agnosia. These findings may be due to an accumulation of clot within the Sylvian fissure, in the frontal lobe, or in the temporal lobe, as well as the subarachnoid space.

Ruptured or unruptured aneurysms of the anterior communicating artery most often produce no focal signs. When sufficiently large they may cause a bitemporal hemianopsia and signs of pituitary insufficiency. When ruptured, they may produce an intracerebral hemorrhage in either frontal lobe with resultant symptoms and signs similar to those caused by a ruptured middle cerebral aneurysm.

Arteriospasm of greater or lesser degree is always associated with the rupture of an intracranial aneurysm. It is usually local in the region of the aneurysm, but it is frequently wide-spread and extends proximally into the internal carotid artery even into the neck and into the entire distribution of the anterior and middle cerebral arteries. As a result of the arteriospasm, there is a diminution in the blood supply which may become so severe as to cause encephalomalacia. Hence, it can cause the same neurological signs which a focal accumulation of clot can produce.

The laboratory studies are not greatly helpful except when hemorrhage has occurred. The cerebrospinal fluid will then contain red blood cells and the pressure will be elevated in three-fourths of the patients. Xanthochromia appears in four hours and may last for four weeks. At first the white blood cells are of the same proportion as that of blood, then polymorphs predominate but by the 5th day there are a greater number of lymphocytes. The total protein is elevated 15 mg. per 10,000 red blood cells per cu. mm. but it becomes more elevated as hemolysis occurs.

X-rays of the skull in children with intracranial aneurysms are usually normal.

Electroencephalography may reveal focal abnormalities and the visual

fields may indicate the size of the lesion. Arteriography is necessary to determine the exact location and the size of the aneurysm, or aneurysms, the degree of the collateral circulation, the extent of the arteriospasm, the presence of an intracranial clot and, hence to indicate the type of therapy. It should be performed as soon as the diagnosis has been made unless the patient is comatose.

The ideal treatment *is the early surgical removal of the aneurysm when possible before hemorrhage occurs. Other surgical methods in order of* effectiveness consist of trapping the aneurysm by clipping the artery on either side of the origin of the aneurysm or by ligation of the internal carotid artery in the neck and intracranially distal to the aneurysm. The neck of the aneurysm itself may be clipped if it is narrow, the aneurysm may be opened, packed with muscle, and the wall cauterized to the muscle to hold it in place, or muscle may be placed around the wall of the aneurysm in order to reinforce the wall. If the direct attack on the aneurysm cannot be accomplished without undue risk to the patient in regard to life itself or increasing the neurological disability, ligation of the internal or perhaps the common carotid artery in the neck can be done. Frequent observations of the patient are made, and if any signs of insufficient collateral circulation appears, the clamp is opened and subsequently closed slowly. The *purpose of the ligation is to so reduce the pressure, the* pulsation, and the jets of arterial blood within the aneurysmal sac that the wall of the aneurysm will contract and clotting will occur within the aneurysm. With organization of the clot the wall will be reinforced so that the aneurysm is no longer a threat to the patient's life.

Progress arteriography about three months postoperatively is necessary to determine the result of therapy. The *prognosis in those cases which* cannot be treated surgically is "extremely grave," as Gowers said, "for no one knows how near or how far may be the fatal rupture." Hence, an heroic attempt should be made to effectively treat them surgically.

Case 6. Aneurysm of Internal Carotid Artery

History: A ten year old boy was well until two months before admission, when he fell down some stairs at school striking his head. He was not unconscious nor did he have any symptoms until two days later when he developed headache in the left frontal and supraorbital regions. This lasted one week, being constantly present and dull in nature. At the end of this week, he developed ptosis on the left eyelid which progressed until it was complete within a few days thereafter. It was noticed that when the lid was lifted, the child had double vision. Because of continuation of symptoms, he was admitted to the hospital.

On examination the positive findings were limited to the complete ptosis on the left side, absence of superior, inferior or medial gaze on the left

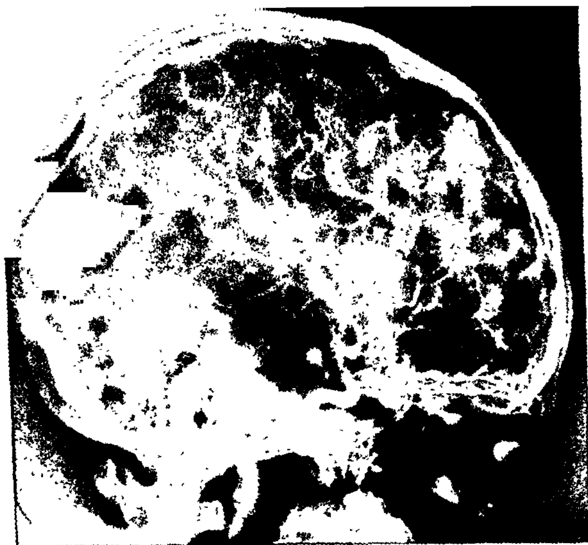


FIGURE 7. (a) An aneurysm of the posterior communicating artery is demonstrated in the carotid arteriogram.

side, and dilatation of the left pupil reacting only slightly to light. X-rays of the skull showed a small epidermoid or eosinophilic granuloma in the left posterior parietal region. A left carotid arteriogram shown in Figure 7 (a) revealed a small saccular aneurysm measuring 7 mm. in diameter arising from the internal carotid in the region of the origin of the posterior communicating artery. The right carotid arteriogram was normal. Carotid compression for thirty minutes caused no evidence of insufficient collateral circulation. The left common carotid artery was ligated by means of a tantalum clip. Two days after operation the patient was able to slightly open the left eye, and five days later he was able to move his eyes medially to some extent. The left internal carotid arteriogram shown in Figure 7 (b) was again performed eleven days later demonstrating all of its intracranial branches. The internal carotid artery was only two-thirds of the caliber it had been eleven days earlier. The aneurysm was no longer visualized. He was last seen four years after operation and was playing

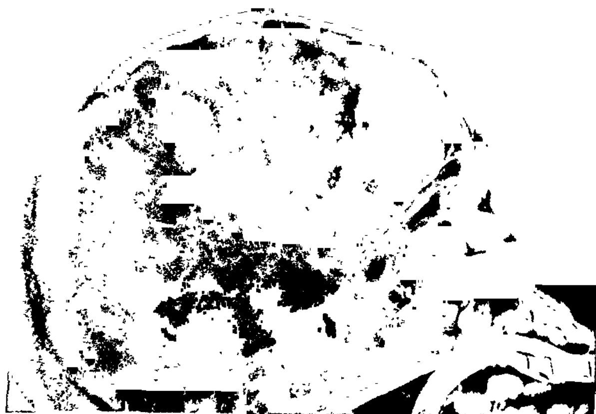


FIGURE 7. (b) The postoperative arteriogram of same patient as in Figure 7a performed eleven days after ligation of the common carotid artery no longer reveals an aneurysm.

football and other sports without difficulty. He did have double vision on downward gaze and some paresis of the inferior rectus muscle. The left palpebral fissure was 2 mm. less in width than the right. The pupils were equal, but the left reacted to light slightly slower than the right.

Spontaneous Subarachnoid Hemorrhage

Spontaneous subarachnoid hemorrhage is a non-traumatic extravasation of blood into the subarachnoid space. It is an episode in the course of an intracranial process and is not a disease entity. The etiology of the hemorrhage is most commonly the rupture of an intracranial aneurysm (80%) or vascular anomaly (10%) but it may be the result of bleeding from a brain tumor, a blood dyscrasia, intracranial venous thrombosis, Hodgkin's disease, acute inflammatory diseases of the brain or acute glomerulonephritis.

It is characterized by the sudden onset of severe headache, vomiting, collapse, and in many instances coma. Examination may show nothing other than stiffness of the neck, a positive Kernig's sign, and blood in the cerebrospinal fluid. The most common focal neurological signs and symptoms are pain in one eye or the forehead, ptosis, dilatation of the pupil

and extraocular muscle weakness which result in diplopia. These symptoms and signs, of course, are the result of hemorrhage from or pressure of an aneurysm of the circle of Willis with involvement of the ophthalmic branch of the 5th cranial nerve and the 3rd, 4th, and 6th cranial nerves. Other signs and symptoms are again dependent on the location of the hemorrhage in the brain. There may be mental changes, convulsive seizures or abnormalities in strength, coordination, reflexes, sensation, or of any other cranial nerves. A bruit occasionally may be heard.

X-rays of the skull in such children are usually normal. The electroencephalogram may show a focal abnormality and the visual fields some defect indicating the side of the lesion. Relief of the headache by percutaneous occlusion of the carotid vessels on one side may give an indication of the side of the lesion. However, none of the preceding signs, symptoms, or special studies are sufficient to identify the etiology and consequently the type of therapy required, unless, of course, a blood dyscrasia, Hodgkin's Disease, acute inflammatory disease of the brain, or acute glomerulonephritis can be diagnosed. Arteriography is indicated and is necessary. Therapy can then be instituted with the knowledge of the exact site of bleeding and the type and location of pathological process which caused the bleeding. Furthermore, it may demonstrate the presence of multiple lesions, the collateral circulation, the presence of an associated intracerebral or subdural clot, other vascular lesions, arteriospasm, or hydrocephalus. If an aneurysm or a vascular abnormality is the cause, surgical treatment is indicated whenever possible, that is, if the location of the pathological process is such that it can be removed, trapped, or clipped intracranially or in the neck, as described previously in this chapter. Hypothermia is used when an aneurysm is approached intracranially. The prognosis, of course, depends upon the damage which has occurred as a result of the hemorrhage and whether or not the underlying pathological process can be effectively treated. (See Case 3.)

Spontaneous Intracerebral Hemorrhage

Non-traumatic intracerebral hemorrhage may be the result of the rupture of a vascular anomaly, of arterial aneurysm or of an arteriosclerotic artery. It may also be due to bleeding from a cerebral neoplasm or secondary to blood dyscrasias, such as hemophilia, sickle cell anemia, thrombocytopenia purpura, leukemia, etc. The most frequent vascular anomalies which rupture and bleed are arteriovenous or capillary angiomas; however, a venous angioma may rarely do so.

The clinical findings are the rather sudden onset of headache and dizziness followed later by vomiting, collapse, and occasionally coma. When the bleeding occurs more slowly, the symptoms gradually appear and

progress over a twenty-four or forty-eight hour period. Once again, other symptoms and signs are dependent upon the area of the brain involved. Since the frontal lobe is the largest, it is the most frequently affected. Therefore, focal seizures, increasing hemiparesis, mental disturbances, hyperactive or reduced reflexes and a positive Babinski sign are common findings. Sensory disturbances, hemianopsias, or aphasias, may occur.

In the differential diagnosis, cerebral thrombosis must be considered and can readily be differentiated by arteriography. The latter will also demonstrate an arteriovenous or venous angioma, but most likely will not show a capillary angioma. The capillary angioma may be diagnosed only by microscopic examination of tissue removed. The blood dyscrasias may be differentiated by the peripheral blood and platelet counts, and the bleeding, coagulation and prothrombin times.

The treatment is the evacuation of the hematoma preferably by craniotomy preceded by arteriography. However, a fresh blood clot may be partially removed by using a large aspirating needle if the child's condition does not warrant the craniotomy. Such a method precludes the identification of the site from which the bleeding has occurred and from which further bleeding may take place.

The prognosis is good when the pathological process can be effectively treated.

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CHAPTER XII

Tumors—Intracranial and Cranial

LYLE FRENCH

THERE ARE special problems related to the diagnosis and treatment of children with brain tumors. These pertain: (1) to the general differences between the care of the adult and pediatric patient; (2) to the characteristics peculiar to tumors occurring in the two age groups, and (3) to the differences required in surgical technique. At first glance one might not think these are especially important. But with a more perspective approach these differences gain in significance. For these reasons an attempt is made in this chapter to delineate anything that might be of help in caring for these patients. Previous reports relating specifically to brain tumors in children include the monograph written by Bailey, Bucy, and Buchanan in 1939 and that by Cuneo and Rand in 1955. Other excellent publications concerned with neurological problems in children, including brain tumors, have been written by Ford, the third edition of whose book was published in 1952 and by Ingraham and Matson, in 1954.

Incidence

The frequency of brain tumors in children is similar to that in adults, i.e., they represent approximately 15 to 20% of brain tumors occurring irrespective of age. This relative frequency was suggested first by Tooth who analyzed a series of 500 patients with brain tumors admitted to the National Hospital in London during the decade of 1902 to 1911. He found 27.8% had occurred in individuals less than twenty-one years of age. From 1911 to 1925 in the same hospital there were 1,008 patients with intracranial tumors of which 12% were in children up to sixteen years of age (Critchley, 1925). Others (Gowers, Bruns, Dowman and Smith, Cushing, 1927) have found an equal frequency. The significant fact emphasized by this incidence is that the possibility of an intracranial neoplasm must be considered strongly in the differential diagnosis of intracranial lesions in a child. Furthermore it has been found that for practical considerations, brain tumors occur with about equal frequency each year of childhood. It is emphasized that this age distribution is for practical considerations and not absolutely correct. Although Schatzke *et al.* have re-

ported on the existence of a brain tumor in a newborn infant and others have reported tumors in infants, there is a lesser frequency during the first year of life. Thereafter the frequency increases up to the fifth to the eighth year due to the greater frequency of medulloblastomas at that time. It then levels off for succeeding years. Figure 1 shows the age frequency in a series of 1,805 verified brain tumors collected from the literature.

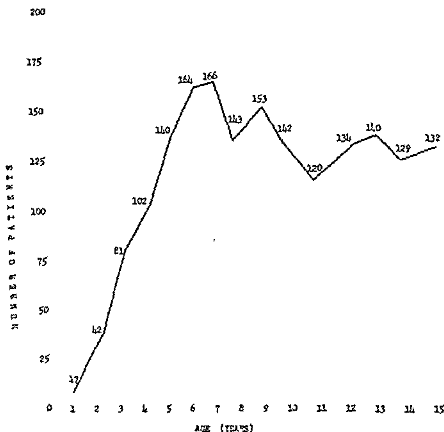


FIGURE 1. Age frequency of brain tumors in children from 1805 cases collected from the literature.

ture. It is apparent that one must be cognizant of the possibility of the presence of a brain tumor irrespective of the age of the child.

Most reports in the literature indicate that the incidence of brain tumors is slightly higher in males than in females. In the series from the University of Minnesota Hospitals the ratio was 146 males to 127 females, not a statistically valid difference. Certain tumor types, however, do seem to be more frequent in males. Medulloblastomas occur almost exclusively in males and the same is true for craniopharyngiomas (*Bailey, Ford*).

Location

The location of brain tumors occurring in children differs from adults since in children a much greater percentage arises in the *infratentorial*

region. This was suggested by Starr and subsequently others have confirmed it. In the series presented here, 131 (48%) were supratentorial and 142 (52%) were infratentorial. In the large series of Ingraham and Matson, 60% were located in the infratentorial region. The important factors producing the variation in the ratio from that of adults are: (1) the infrequency in children of meningeal tumors which in adults occur predominantly in the supratentorial region, particularly since this group comprises about 20% of the total brain tumors in adults; (2) the fact that astrocytomas have approximately the same incidence in children as in adults, but in children they usually are located in the posterior fossa, and (3) the occurrence of medulloblastomas in the infratentorial region of children.

Histology

Figure 2 shows the frequency of brain tumors in children according to histological types. The infrequency of meningiomas is apparent. An interesting fact is that when meningiomas do occur, they often are part of von Recklinghausen's disease and tend to be much more malignant than those occurring in adults. Neuromas which are primarily acoustic neuromas, occur slightly less frequently than in adults; in children they often are bilateral and again are part of von Recklinghausen's disease. Gliomas comprise 71% of the tumors in this series, whereas in adults the usual frequency is about 40%. Ford estimated that gliomas comprise 75% of all intracranial tumors in children, Ingraham and Matson found 80% to be gliomas, and Cuneo and Rand found 70% to be gliomas. In the glioma group, medulloblastomas are frequent, a tumor type that seldom if ever occurs in an individual over twenty years of age. The glioblastomas are less frequent than in adults and when they do occur are often located in the pons or medulla. The astrocytomas tend to be located in the posterior fossa and not infrequently they have a characteristic large cyst with a small mural nodule, the cystic type astrocytomas, a tumor type seldom found in adults. The infrequency of pituitary tumors in children found in this series has been observed by many reporters.

The tumors occurring in infants and younger children tend to lie along the midline (chiasmal region, third ventricle, brain stem), those in older children in the cerebral or cerebellar hemispheres.

Signs and Symptoms

What are the symptoms produced by an intracranial neoplasm in a child? Much depends upon the age of the child and upon the location of the tumor. In general, the symptoms can be divided into two main groups, those due to *increased intracranial pressure* and those due to *local involvement of the brain* either by the tumor or by the associated cerebral edema.

The first symptoms observed may result from either of these two factors. It is amazing how few and seemingly insignificant symptoms may be present even though evidence of extreme pressure is found on ophthalmoscopic examination, plain skull roentgenograms, or air studies. Needless to say it is best if a tumor can be suspected prior to the development of such pressure. This can be done only by being constantly alert to the problem. No suggestive symptoms should be shrugged off as insignificant. It is fortunate there are certain fairly constant symptoms that occur in children with brain tumors which lead one to suspect this diagnosis. Only by constant vigilance, however, can the more occult tumors be discerned.

Tumor Type	*% of Gliomas*		*% of Total Tumors*			*Number of Brain Tumors*		
Neuroma					1			4
Meningioma					3			7
Glioma					71			195
Astrocytoma		37		27			73	
Astroblastoma		3		2			6	
Glioblastoma		13		10			26	
Oligodendroglioma		3		2			5	
Spongioblastoma		3		2			5	
Medulloblastoma		15		11			29	
Ependymoma		16		11			32	
Papillomatous	2		1			4		
Myxopapillary	2		2			5		
Epithelial	5		4			10		
Cellular	7		4			13		
Ependymoblastoma		5		2			8	
Pinealoma		5		4			10	
Ganglioneuroma		1		*			1	
Vascular (angioblastoma)					4			11
Mixed Tissue					3			7
Epidermoid				1			4	
Dermoid				*			1	
Teratoma				*			2	
Hypophyseal					5			
Craniopharyngioma				4			11	12
Chromophobe				*			1	
Granuloma					3			9
Metastatic					8			22
Miscellaneous					2			6
Cyst				*			1	
Fibroblastoma				1			3	
Sarcoma				1			2	
								273

* Less than 1%

FIGURE 2. Frequency of 273 brain tumors in children according to histological type.

When a child suspected of harboring an intracranial lesion is examined, emphasis should be put on the special techniques and features that are peculiar to children. There are too many to enumerate but included in this list are the normal persistence of a Babinski response up to eighteen months of age, the utilization of a child's natural imitative tendencies in getting cooperation for such tests as finger-to-nose, etc., observation of the child at play, notation of any change in handedness, deterioration in school performance, and aplasia resulting from involvement of the pyramidal tract in growing infants and children. Even so, examination of a child *suspected of having a brain tumor may reveal only signs of increased* intracranial pressure. This is especially true early. Evidence of such increased pressure are papilledema, dilatation of the veins in the skin over the frontal and occipital regions, widening of the cranial sutures, bulging of the anterior fontanelle, increased head circumference, and strabismus. Roentgenographic evidence of increased pressure is present in approximately 90% of cases.

A staggering gait or a tendency to stand and walk with a wide base may be an early indication of an intracranial lesion. Clumsiness with one hand may be noted by the parents. While these are generally considered to be localizing signs, in some children they occur early and alert one to the

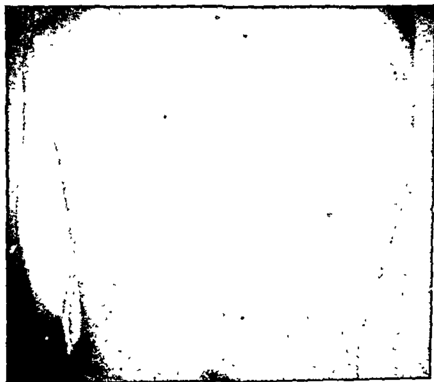


FIGURE 3. Child with multiple neurofibromatosis. Café au lait spots typical of this lesion are visible. This patient had a right temporal lobe glioblastoma multiforme.

presence of a neoplasm. The same is true for abnormal body growth.

The presence of congenital deformities, such as multiple areas of cutaneous pigmentation (café au lait spots), absence of the roof of the orbit, syndactylism, or asymmetrical physical development should bring to mind the possible presence of an associated intracranial tumor. Dimples, sinus tracts, and abnormal hairs may signify an intracranial lesion, probably a mixed tissue tumor. The presence of a bruit over the skull is highly suggestive of an intracranial vascular malformation. However, such bruits have been observed in children with cervical adenitis which has produced secondary pressure or inflammatory involvement of the carotid and occipital vessels.

Disturbance in Personality

A change in personality constitutes one of the earliest symptoms and occurs at some time during the illness in almost one-half of all the children with brain tumors. This change is found as frequently in patients with supratentorial tumors as in those with infratentorial tumors. Because of the insidious onset, these early symptoms frequently are not noticed. The child initially loses interest, especially in playing with other children. There is loss of appetite and a preference to "go lie down" rather than to eat or play. He is more easily fatigued; developmental progress is retarded; and he may become weak and listless. There is a tendency to enter less into the activities of school. Some children become restless, more irritable, and cry or complain readily. Reports in the literature concerning the comparative mental reactions in children with cerebral and cerebellar tumors state that the latter are good natured, cooperative and alert in spite of severe headaches, nausea, and vomiting. Conversely, children with supratentorial tumors have been considered to be more irritable and uncooperative. There is considerable doubt that this differentiation actually exists. More probably, the frequency and severity of mental reactions are comparable in the two groups.

Headache

Obviously the younger the child the less the symptoms are verbalized, so that many complaints common in adults are overlooked in children. Unfortunately these often are symptoms which, if recognized, could lead to earlier diagnosis. Headaches fall into this category. Sometimes the only evidence of a headache is increased irritability. The headache in the majority of children is not specifically located. Some children do signify the location of headache by holding their hands over the affected area, such as above the eyes or the occipital region. The location of the headache

may be significant of the location of the tumor. Pressure by the tumor or by edematous brain tissue on the falx, superior surface of the tentorium, or blood vessels, all of which are innervated by the first division of the trigeminal nerve causes pain to be referred to the cutaneous distribution of this nerve, i.e., the eye and forehead. Headaches occurring over the occipital or posterior cervical region are due to irritation of the ninth and tenth cranial nerves and the posterior roots of the cervical nerves which supply the dura in the region of the foramen magnum. This irritation produces the nuchal rigidity, simulating meningitis, often seen in children with posterior fossa neoplasms. It also explains the tendency for children with posterior fossa tumors to hold their heads tilted to one side. Any increase in the intracranial pressure due to coughing, sneezing, or straining may initiate headaches or occipital pains by traction on these pain-sensitive structures. Tenderness may be present over the suboccipital area. Headaches, accompanied by vomiting, frequently occur in the morning after the child has arisen from bed and are due to a change in dynamics of the blood and cerebrospinal fluid pressure when the child assumes an upright position. Following the vomiting the headache is often relieved. Another characteristic type of headache is one that comes on when the child assumes an upright position and is relieved when he bends over in a knee-chest position. Children usually are quick to find this method of relief and at-

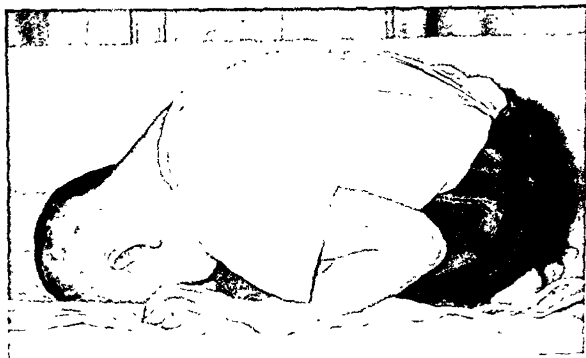


FIGURE 4. Child with craniopharyngioma—position assumed to relieve headaches. In this position the tumor moves forward permitting proper flow of cerebrospinal fluid through the third ventricle.

tempt to maintain the knee-chest position at all times (Figure 4). This type of headache may be observed in patients with intraventricular tumors, such as papillomas, pinealomas (Nash, Harris), and with craniopharyngiomas. In these instances the tumor acts as a movable valve that may occlude the aqueduct of Sylvius or foramen of Monro, thereby producing a rather sudden obstruction to the flow of cerebrospinal fluid. This sudden increase in the supratentorial tension may result in a herniation of the medial portion of the temporal lobe through the incisura of the tentorium, thus compressing the structures normally passing through the foramen. Symptoms and signs generally considered falsely-localizing may result. For example, compression of the posterior cerebral artery by the free edge of the tentorium may produce an homonymous hemianopsia, compression of the oculomotor nerve at the same site may result in ptosis, dilated pupil, and strabismus. The long tracts, either motor or sensory, can be involved by compression of the pes pedunculi by the incisura. This compression may even result in death from edema or hemorrhage in the midbrain. Another site at which herniation may occur is the foramen magnum. The tonsillar portion of the cerebellar hemispheres may be pushed downward through the foramen. This is more apt to occur in patients with space occupying lesions in the posterior fossa. Symptoms produced are due to compression of the upper cervical nerves, the lower cranial nerves, or of the medulla. There may be changes in vital signs, nuchal rigidity, tilting of the head, loss of conjugate ocular movements, etc.

Vomiting

Vomiting is the second most frequent symptom in children with brain tumors. It occurs irrespective of the type or location of the tumor. Critchley (1928) found it present in 72% and Globus *et al.* (1942) in 73% of all children with brain tumors. However, in the author's experience, vomiting alone was the initial symptom in only 10% of these children; but vomiting associated with some other complaint such as headache, nausea, or stiff neck has been the initial symptom in 33%. Vomiting occurs at some time during the course of the illness in approximately 80% of the children. Vomiting from intracranial tumors may be associated with abdominal pains and this combination of symptoms may simulate appendicitis. An appendectomy may be done and only when no relief ensues does the physician recognize the possibility of a brain tumor being present. Likewise tonsillectomies have been done to relieve vomiting which later was found due to a brain tumor.

Vomiting may occur over a protracted period of time and be so severe that the child becomes dehydrated to the extent that the intracranial pressure does not become elevated even though the child has a very large

intracranial tumor. The anterior fontanelle may remain scaphoid or depressed, the head circumference may not be enlarged. None of the usual signs suggestive of an intracranial neoplasm may be evident.

Often there is a certain intermittency of headache and vomiting, especially in children under seven or eight years of age. Critchley believed this fluctuation of symptoms signified the presence of a tuberculoma. An alternate and more logical explanation is that symptoms produced by increased intracranial pressure are periodically relieved by a progressive widening of the cranial sutures (Bailey *et al.* 1939) or by an intermittent recession of cerebral edema.

Nausea

Nausea is an infrequent initial symptom in children with brain tumors, although it occurs at some time during the course of the illness in about one-fifth of these patients. The reason for its infrequency is the same as for headaches—children, especially younger ones, do not verbalize their complaints.

Disturbance in Vision

Impaired vision is an unusual initial symptom in children with brain tumors. When present it often is first noticed by the school teacher. Visual loss can be due to one or more of the following four conditions: (1) *Papilledema and secondary optic atrophy;* (2) *primary optic atrophy;* (3) *visual field defects,* and (4) *diplopia.*

Papilledema is due to increased intracranial pressure, but many children have increased pressure without ever developing papilledema. Gradual compensation may be made to the space occupying lesion by progressive enlargement of the head which occurs as a result of widening of the cranial sutures. Another reason for lack of papilledema in the presence of an intracranial neoplasm is the existence of a developmental variation in the extent that the subarachnoid space projects out along the optic nerve. If there is a short projection the retinal veins may not cross the space, so that an increased intracranial pressure cannot embarrass the venous return from the retina. When papilledema is present it is highly significant. It is an absolute indication for immediate investigation. If for some reason the procedure designed to relieve the increased pressure, as evidence by the papilledema, cannot be carried out soon daily checks on the visual acuity are mandatory. Children can change from slightly diminished vision to total loss within a few days and when total loss occurs it may not return even though the pressure is relieved. If the increased pressure is of long standing, the only fundiscopic evidence may be secondary optic atrophy. An item to be remembered in children with recurrent tumors and increased intracranial pressure is that papilledema may not occur if an optic atrophy

is present because the gliosis in the optic nerve prevents the development of the edema.

Primary optic atrophy occurs from direct pressure on the nerve. In children the most usual cause is a craniopharyngioma but other tumors in this region will also produce a primary atrophy. There may be an associated *visual field defect.* This combination of field defect and primary optic atrophy is almost pathognomonic of a lesion in the region of the hypophysis. Field defects may occur also with neoplasms located in the parietal, temporal, or occipital region but in these instances the optic atrophy is not of a primary type but rather secondary to an associated papilledema. Collier described an hemianopsia occurring as a false-localizing sign due to compression of the posterior cerebral artery by the incisura of the tentorium. Learmouth *et al.* reported bilateral cecocentral scotomas and bitemporal hemianopsia for color occurring in a patient with a posterior fossa neoplasm. Wagener and Cusick reported a similar case and stated that in these instances the visual complaint and field defect occur late in the course of the illness and are due to pressure on the optic pathways by a dilated third ventricle. Others (Dandy (1933), Bailey (1924), Courville (1928), Jefferson and Jackson) have reported similar cases. A posterior fossa tumor producing a dilated third ventricle may also produce pressure on the surrounding hypothalamic region, causing hypersomnia, polydipsia, and polyuria. Care must be taken in interpreting any sign that occurs after the development of increased intracranial pressure for, like those above, it may be falsely localizing.

Diplopia occurs in at least one-fourth of children with brain tumors. It is usually due to involvement of the third or the sixth cranial nerve. Perhaps the fourth cranial nerve is as frequently involved but more difficult to discern. The exact mechanism producing these palsies is not always the same; often they are due simply to increased intracranial pressure and may be of no specific localizing value.

Nystagmus

The nystagmus that is of most significance in localization of brain tumors in children is a nystagmus of fixation (Bailey *et al.* 1939) which appears only when the gaze is fixed on some object. Fundamentally, it is dependent on an imbalance of involvement of the vestibular centers on the two sides. An irritative lesion on one side will produce either a forced deviation of the eyes toward the other side or nystagmus with the quick component toward the irritated side. A destructive lesion, of course, produces deviation of the eyes toward the side of involvement or nystagmus with the quick component to the opposite side. Unilateral cerebellar tumors (astrocytomas) may do this, but unfortunately even with seemingly unilateral tumors there actually may be bilateral involvement with corresponding

bilateral vestibular dysfunction. In this event the components on looking in either direction may be equal to each other or there may be no nystagmus. However, if definite coarse movements are obtained on deviation of the eyes in one direction, and fine rapid movements or no nystagmus on looking in the other direction, the lesion is on the side of the direction of gaze producing coarse nystagmus in about 80% of the cases. Medulloblastomas may not produce nystagmus because of bilateral involvement of the vestibular centers as suggested above. If the nystagmus does occur in a patient with a medulloblastoma, it indicates more involvement on one side than the other. Nystagmus is a sign not only of cerebellar lesions but also occurs in about one-half of the patients with pontine and pineal tumors (Horrax and Bailey, French 1947). A vertical nystagmus is indicative of lesions in the region of the superior vestibular nucleus (Kahn *et al.*). In patients with unilateral cerebellar hemisphere involvement (astrocytoma) the eyes at rest may tend to deviate slightly toward the side opposite the lesion.

Cranial Nerve Involvement

Involvement of the cranial nerves, other than the extra-ocular nerves, occurs in about one-fifth of children with brain tumors, but the only location in which involvement is of sufficient frequency to be of diagnostic value is in brain stem tumors. Tumors in this location become manifest early, and often the only sign for long periods of time is involvement of some of the last six cranial nerves (Foerster and Gagel 1940, Horrax and Bailey).

Parinaud's Syndrome

Restricted upward gaze is considered due to pressure on the collicular region of the midbrain. Dandy (1933) has stated that the first sign of restricted upward gaze is ptosis; when present bilaterally without further extra-ocular muscle palsies, he believes this finding is pathognomonic of a tumor pressing down from the region of the pineal gland. Ptosis is present in 80% of the patients with pineal tumors, but also may occur in patients with tumors of the superior vermis, of the pontine region, or of the third ventricle region. Tumors so located can exert pressure on, or involve by direct infiltration, the tectum of the midbrain which is considered the center for upward gaze

Rhinorrhea

An unusual symptom is the spontaneous escape of cerebrospinal fluid from the nose. In 1926 Locke reviewed this subject and concluded that obstruction to the flow of cerebrospinal fluid with increased intracranial pressure was necessary in the formation of a fistula of this type. In addition

to tumors, rhinorrhea has been reported in a patient with an Arnold-Chiari deformity (Youngs and Peyton).

Ataxia

Ataxia involving primarily the legs or trunk and manifested by an unsteady gait with a wide base and a positive Romberg sign is present in 60% of the patients with cerebellar lesions. It is due to involvement of the vermis and the flocculonodular lobe. Incoordination (adiadokokinesis, past pointing, intention tremor, rebound) in the upper extremities signifies involvement of the cerebellar hemispheres. Often both areas are involved either directly by tumor or indirectly by pressure from the tumor making differential localization impossible. According to Marburg (1942) the ataxia occasionally seen in patients with supratentorial tumors may be due to pressure through the tentorium onto the cerebellum. Bruns emphasized the importance of what he called "d'ataxie statique" of frontal lobe tumors. He called attention to the fact that the sequence in which sympoms appear is essential to differentiate frontal lobe from cerebellar tumors. Frontal lobe tumors produce motor weakness early whereas cerebellar lesions produce ataxia early. This simply re-emphasizes the known fact that symptoms and signs occurring early in the course of the illness usually are of much more value in localization than those occurring later. In fact, localizing symptoms and signs appearing late in the course of the illness, when general signs alone have pre-existed, are often falsely-localizing due to the occurrence of cerebral edema.

Grant emphasized the possibility of cerebellar symptoms being produced by supratentorial tumors, and stated that a sagging tentorium from occipital, occipitoparietal, or thalamic tumors may give sufficient mechanical cause for cerebellar dysfunction. Likewise pineal tumors may produce incoordination by direct invasion of or pressure on the brachium conjunctivum.

Paresis

A true paresis of an extremity occurs rarely in children with cerebellar tumors, but is present in about one-third of those with cerebral tumors and in about one-half of those with brain stem tumors. A cerebellar tumor can produce a true paresis only if it is large enough to produce pressure on or directly infiltrate into the brain stem. In the latter instance it is, of course, no longer specifically a cerebellar tumor.

Abnormalities in Tendon Reflexes

Abnormalities in the tendon reflexes are found in about 85% of children with brain tumors, and are about three times more frequent in those with

supratentorial than with infratentorial tumors. Hyperactive reflexes are highly indicative of a supratentorial lesion but may occur in the presence of an infratentorial tumor. If the hyperactivity is unilateral, the lesion is most apt to be supratentorial. However, unilaterally hyperactive reflexes may occur with either midline or hemispheric cerebellar tumors, so that the occurrence of this abnormality cannot be used diagnostically to eliminate a cerebellar tumor. Bilaterally hyperactive reflexes may occur in patients with either a supratentorial or infratentorial lesion. Hypoactivity of reflexes and hypotonia (so called pendular reflexes) are classical of cerebellar lesions. These may be bilateral or unilateral depending on whether the area of the vermis or the cerebellar hemispheres are involved.

Babinski Sign

Bilaterally positive toe signs (Babinski) are of no localizing value, for they may be present irrespective of location of the tumor and are normally present in infants up to 18 months of age. Unilaterally positive toe signs may be of localizing significance, for they indicate unilateral motor tract involvement.

Convulsive Seizures

Generalized convulsive seizures in children may be produced by a great variety of conditions and hence are of relatively little value in the diagnosis of intracranial tumors. Approximately 30% of children with cerebral tumors and 5% of those with cerebellar tumors have generalized seizures. They have been observed to occur in patients with tumors in any one of the various regions of the brain. Jacksonian type seizures occur mainly in patients with cerebral tumors but also may be present in those with cerebellar lesions. The author almost missed a benign cystic astrocytoma of the cerebellum when he questioned the advisability of performing a ventriculostomy in a five-year-old girl who appeared moribund and had as her only known symptom focal convulsive seizures in the left hand. Fortunately the ventriculostomy was done (as a last resort!) and a hydrocephalic ventricle encountered. The child perked up and two days later the cerebellar tumor was totally removed. The child is now symptom free, five years after the operation. There is no absolute nor infallible symptom or sign! Jacksonian seizures in the presence of the cerebellar neoplasm are usually explained by the occurrence of an implantation of the cerebellar lesion in the supratentorial region. An alternate explanation is cortical irritation on the basis of transmitted pressure (Courville). There is no doubt that any one of these explanations may be the "modus operandi" "Cerebellar fits" are not true convulsive seizures but are temporary attacks of decerebration. They may occur, as their name implies, in patients with cerebellar

tumors, are very infrequent, and are probably due to interruption, through pressure, of the efferent stimuli from the anterior lobe of the cerebellum. Frequently they are accompanied by cyanosis, respiratory irregularities, and unconsciousness (Bailey *et al.* 1939).

Diabetes Insipidus

Polyuria, polydipsia, and abnormalities in temperature regulation are symptoms observed in patients with tumors so placed that pressure is exerted on the hypothalamic nuclei or in whom there is direct infiltration by the tumor of the hypothalamus. These symptoms have been observed in this series in 30% of patients with pineal tumors and in 40% of patients with tumors of the hypophyseal region. There are reports in the literature (Dandy, Cushing 1923, Bailey) that such symptoms may also be produced by a dilated third ventricle in the presence of a posterior fossa neoplasm.

Abnormal Body Growth

A Fröhlich type of dystrophy is unusual in children with brain tumors (Worster-Drought *et al.*, von Bogaert), and when it occurs, it is indicative of involvement of the hypothalamic region. Fröhlich's syndrome has been observed in children with pineal tumors primarily in the third ventricular region. Gigantism may occur in older children with pituitary tumors.

Occiput Tenderness

Tenderness over the occiput may be found in children with posterior fossa tumors. Usually this tenderness is unilateral, and is an excellent confirmatory sign of an underlying tumor.

Diagnostic Procedures

Even though the presence of an intracranial neoplasm is unquestioned, the exact location may not be so evident. At times it is quite impossible by clinical examination to distinguish whether a tumor is supratentorial or infratentorial. Further studies are needed. Routinely these should include skull roentgenograms. The views required are antero-posterior, postero-anterior, basilar, and either stereoscopic laterals or right and left laterals. If a projection or exposure of a film is inadequate it should be repeated. The calcium sometimes present in tumors of children is frequently finely stippled requiring the best radiographic techniques for visualization. The same is true for areas of bone erosion which may be barely distinguishable in the very thin skull of an infant. Special care should be taken to look along the midline, especially in the occipital bone, for small openings which are characteristic of congenital dermal sinuses, mixed tissue tumors, etc. The views should include the upper cervical spine to rule out a platybasia or basilar impression.

If plain roentgenograms are not clarifying, further studies of pneumoencephalography, ventriculography, or angiography are required. The use of pneumoencephalography in a child suspected of harboring a tumor is extremely limited. Only in cases of suspected brain stem gliomas, or acoustic neuromas, or very occasionally after ventriculography has been performed and there is still uncertainty of diagnosis, is pneumoencephalography permissable. In other instances, the dangers far outweigh any possible advantages. Pressure changes that produce complicating cerebral edema or actual shift of parts of the brain or of the tumor itself may result in obstruction of the few remaining open channels for the flow of cerebrospinal fluid. Because one might "get away with it" is no reason to jeopardize the life of the patient. There is no place for shortcuts.

Roentgenographic Features

Three radiographic abnormalities frequently observed in children with increased intracranial pressure are: (1) widening of the cranial sutures with enlargement in circumference of the head; (2) increased convolutional

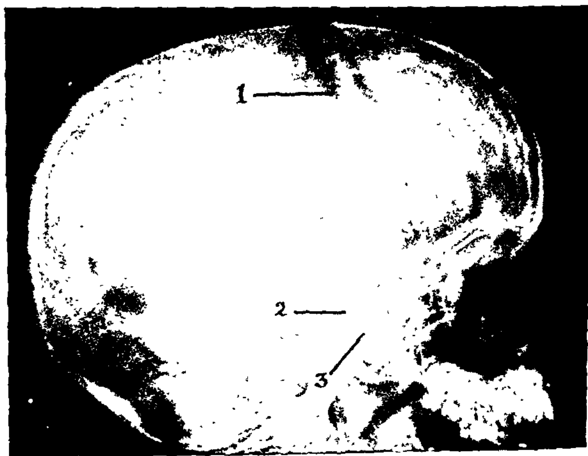


FIGURE 5 Roentgenogram showing evidence of increased intracranial pressure. 1. Widening of the cranial sutures 2. Erosion of the posterior clinoids 3. A slightly decalcified sella

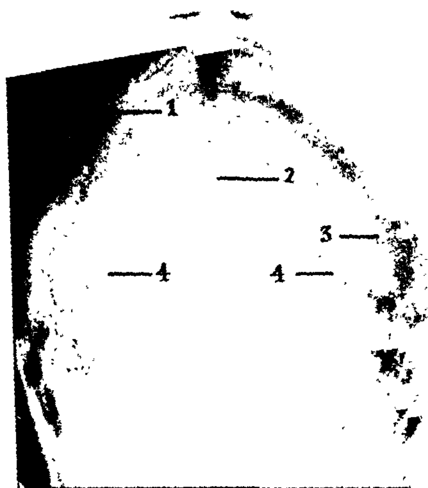


FIGURE 6. Roentgenogram of a child with a left cerebellar astrocytoma showing evidence of increased intracranial pressure. Numbers 1 and 3 *identify* respectively the Lambdoid and Occipitomastoid Sutures, which are not widened. Numbers 2 and 4 identify respectively the sagittal and coronal sutures, which are widened.

(digital) markings; and (3) erosion or decalcification of the dorsum sellae. Whereas any one of these three abnormalities may justify a conclusion of increased intracranial pressure, it is preferable that two of the three be present. Eighty per cent of children with brain tumors reveal, according to these criteria, roentgenographic evidence of increased intracranial pressure. In children under five years of age, increased pressure is more frequently accompanied by widening of the cranial sutures. In children over ten years of age, widening of the more firmly united sutures is less frequent but erosion or decalcification of the dorsum sellae is often seen.

Local erosion of that portion of the calvarium immediately overlying a tumor should not be confused with the erosion of increased convolutional markings which is due to a generalized increase in intracranial pressure. Whereas this local erosion is infrequently observed, when it does occur

in children it is an accurate indication of an underlying tumor, e.g., unilateral erosion of the occipital bone in patients with cerebellar astrocytomas.

In 1925, Naffziger reported that a shift in the position of the pineal gland may be of great value in localizing brain tumors. However, the pineal gland is seldom calcified until late in childhood, making this method of localization of less value in children than in older individuals. Dyke in 1930, Fray in 1938, and Hawes and Mead in 1943, reported that a posterior displacement of the pineal gland may sometimes occur in patients with infratentorial tumors. This is just the reverse of the more frequently seen upward shift (Vaestine and Kinney) and is believed to be due to an internal hydrocephalus resulting from a small obstructing infratentorial lesion. However, evidence of any shift in the pineal gland is very unusual in children with infratentorial tumors.

In 1925 van Dessel reported that calcium may occur in any slowly growing intracranial tumor. He also stated that there was a greater frequency of calcification in tumors of older people than in tumors in children. However, Boldrey and Miller have reported the presence of calcium in rapidly growing tumors of very young children. Actually there is roentgenographic evidence of calcification in about 15 to 20% of tumors in children.

Skull defects due to invasion by metastatic tumors may occur in children. A tumor which is characterized by metastases primarily to the skull is Hutchinson's type of neuroblastoma of the adrenal gland.

Angiography

The question then arises whether ventriculography or angiography is advisable. The answer is best determined by the experience of the surgeon. Some no doubt feel that angiography is sufficiently difficult in infants and children to make its use inadvisable. The advantage of angiography is that it does not demand immediate surgical intervention quite as rapidly as ventriculography and may make the technical part of the operation easier by visualizing the vascular system in and adjacent to the tumor. It may also afford knowledge of the histology of the tumor. Recently the method of performing angiography in children in this clinic has been to put the child to sleep with pentothal, to insert an endotracheal tube, and to make an attempt to enter the carotid or vertebral artery percutaneously. If successful, angiography is carried out with the usual techniques. However, if it seems improbable that the vessel will be cannulated easily, a reflux brachial angiogram is performed. This technique has been described by Gould *et al.* and consists of direct visualization and cannulization of the brachial artery in the mid-portion of the upper arm. This can be done either on the right or the left side. About 8 cc. to 20 cc. of 35% diodrast, depending upon the



FIGURE 7. (a) Lateral view. Angiogram done with the reflux brachial technique. Adequate visualization of both the carotid and vertebral systems can be obtained.

size of the child, is then injected refluxly up the brachial artery and quite invariably adequate filling of not only the vertebral-basilar system but also the carotid system is obtained. The latter can be made more certain by compression of the ipsilateral carotid artery. In fact, the technique seems to be accomplished so easily that often no attempt is made to obtain either a vertebral or carotid angiogram by direct percutaneous injection of the respective vessels (Figure 7 a and b). The reflux type angiogram is done as a first measure.

If a lesion in the supratentorial area is visualized and is consistent with the previous clinical assessment measures are taken to carry out a craniotomy. The techniques used are dependent on the location of the lesion and are discussed under the various tumor types.

Ventriculography and Ventriculostomy

If the patient has increased intracranial pressure and the general condition is poor and one feels the lesion is in the posterior fossa there is no

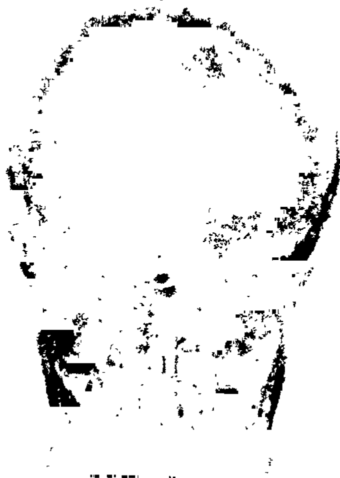


FIGURE 7. (b) AP view. Angiogram done with the reflux brachial technique. Adequate visualization of both the carotid and vertebral systems can be obtained.

question that ventriculography with a *ventriculostomy* tube left in place is the best approach. In fact, if the patient is suspected of harboring a cerebellar neoplasm and is acutely ill at the time of admission ventricular drainage by catheter always is instituted immediately. This procedure was emphasized by Fincher, and makes possible a reduction of cerebrospinal fluid pressure and its maintenance at a normal level. Following the release of intracranial pressure the patient can be examined more adequately. If open drainage of the tube is permitted air will enter as fluid escapes and filling of the ventricular system with air is obtained automatically. If a closed system is employed, air studies can be carried out through the tube after the child's condition has improved sufficiently. After such ventricular drainage in the presence of a posterior fossa neoplasm the patient's condition invariably improves. If it fails to improve a strong suspicion is warranted that the lesion is located supratentorially. An exception to this is the occasion in which a large firm neoplasm fills the fourth ventricle or



FIGURE 8. Ventriculogram of a child with a left cerebellar astrocytoma. Arrows point to elevation of floor of third ventricle. This must be differentiated from the deformity produced by a tumor in the hypothalamus or upper brain stem.

the tumor is located anteriorly in the cerebellar region so that the neoplasm may herniate upward through the incisura resulting in further compression of the brain stem. This, however, is extremely unusual. When it does occur, the prognosis is extremely poor. If it is indefinite whether the tumor is located above or below the tentorium, the ventriculostomy tube, or the ventricular needle at the time of ventriculography, always should be placed in the ventricle on the side of the suspected lesion. The reason for this is that the ventricle on the side of the tumor often is small whereas the contralateral ventricle usually is enlarged. Tapping of the enlarged ventricle may permit sufficient shift and collapse of the smaller ventricle that later insertion of the needle on that side is impossible. If an estimate of ventricular size is made by determining the depth at which the ventricle is encountered, a dilated ventrical contralateral to a supratentorial neoplasm can be mistaken for a hydrocephalus associated with an infratentorial lesion. This mistake has been made by the author with almost fatal consequences. After ventriculography the lateral ventricles, the third ventricle, and the aqueduct may fill and all the system found to be dilated making

the *presence of an infratentorial lesion very apparent.* However, it is not always quite that easy. There may be no filling of the third ventricle or the aqueduct even though the lateral ventricles are very dilated. The question that then arises is whether one is dealing with a lesion in the third ventricle, along the aqueduct, or down in the posterior fossa. Peterson and Baker have emphasized the difficulties of distinguishing these tumors. If it is obvious that the patient does have a lesion but it cannot be ascertained in which of these three sites it is located, it is then preferable to perform *a posterior fossa craniectomy and try to determine if the lesion is in this* region and if not to insert a ventriculo-cisternal catheter (Torkildsen) thereby shunting the flow of cerebrospinal fluid around any possible lesion in the midbrain or the third ventricular region. Before inserting such a tube it is questionable just how much of a posterior fossa exploration is permissible. An extensive exploration rather precludes a functioning shunt. Certainly one cannot get an appreciable amount of blood or necrotic material in the region of the cisterna magna and expect the "Torkildsen" tube to work. *It is believed by many surgeons that visualization of the cerebellar* tonsils to see if they are herniated down into the foramen magnum and inspection to ascertain whether one hemisphere is larger or is more tense to palpation than the other is adequate to determine the presence or absence of a posterior fossa tumor in a child. However, it is believed by the author that prior to calling this a negative exploration it is advisable to look through the foramen of Magendie into the fourth ventricle. To do so one must enlarge the foramen. If no lesion is found, dye, either phenosulfothalein or indigo carmen, should be instilled into the lateral ventricle. The existence of a block can be determined by the failure of the dye to appear at the lower level of the aqueduct. Actually the eventual appearance of the dye does not rule out a block or at least an incomplete block. The rapidity with which the dye comes down must be observed because dye should appear within a few minutes at the lower end of the aqueduct after its instillation into the lateral ventricle. If it doesn't appear in this time it is highly probable that an obstruction lies above that point. The mere presence of excessively large amounts of fluid in the region of the cisterna magna does not preclude a partially obstructing lesion in the third ventricle or aqueduct. This has been emphasized by Kahn *et al.* in their book.

Correlative Neurosurgery

In cases in which *there is considerable doubt whether a neoplasm is* harbored within one of the cerebellar hemispheres it is considered advisable to insert a ventricular needle up into the cerebellar hemispheres hoping that one can either aspirate cystic fluid or tumor tissue. Extreme care must be

taken not to overlook a neoplasm located high up in the posterior fossa near the incisura. If no neoplasm is encountered, an obstructed cerebrospinal fluid flow at the incisura might be secondary to adhesions (meningitis or subarachnoid blood). If no obstruction is seen at the incisura, could the difficulty be a lack of absorption secondary to obliteration of the subarachnoid spaces over the cerebral hemispheres by meningitis or blood? Such adhesive arachnoiditis can usually be visualized at the time of doing the ventriculostomy providing the trepination is made with a perforator and burr rather than with the small drill sometimes advocated. Occasionally one is confronted with a circumstance in which there are dilated lateral ventricles, dilated third ventricle and aqueduct, and when the posterior fossa is exposed, no neoplasm is apparent. What to do now! About the only feasible procedure is a ventriculocisternostomy (Torkildsen). The lower end of the tube classically is placed into the cisterna magna. An alternate location that serves very well, especially if the posterior fossa is mutilated by the usually thorough exploration, is in the cervical region (C_2-C_3). Matson uses this site preferentially and recently the author has found it very satisfactory. The tube is placed anterior to the dentate ligament, into the subarachnoid space anterior to the cord.

An alteration of cerebrospinal fluid dynamics can be very disturbing to a child with increased intracranial pressure. Whenever a ventricle is cannulated either for an air study or to decompress the cranial cavity, the fluid should be released gradually. Too rapid decompression, especially if the ventricles are dilated, may permit collapse of the cerebral mantel with tearing of the small vessels that run from the cortex to the meninges and the development of a subdural hematoma. This complication is especially likely in younger children and infants because of the lesser resilience of the brain.

In older children an attempt always is made to perform the ventriculogram under local anesthesia. Often this is not possible because of lack of cooperation or because of an apparent psychological hazard to the child. When general anesthesia is used, an endotracheal tube always is inserted to assure control of respiration. Because the ventriculogram, even under ideal circumstances, may be very disturbing preparation must always be made to follow it with a craniotomy which may become necessary.

It is the policy when doing a ventricular drainage to use a Robinson type urethral catheter size No. 8 French which has either never been used before or has been used only for an uncontaminated neurological case. These catheters are soft and pliable. They are not apt to straighten out and become imbedded in the ventricular wall. By inserting a spinal needle into the lumen of this catheter as shown in Figure 9 the catheter is sufficiently rigid for insertion. When the catheter and needle are placed into

the ventricle, fluid will come out either through the needle from which the stylet has been removed or through the catheter itself. The needle itself is then removed. Other types of needles have been used but it has been found that a No. 20 spinal needle is the best because it does not break through the soft end of the catheter and because this calibre needle does not completely fill the catheter thus facilitating withdrawal of the needle. When a ventriculogram is obtained, the usual standard projections should be made. In addition, the hanging head projection emphasized by

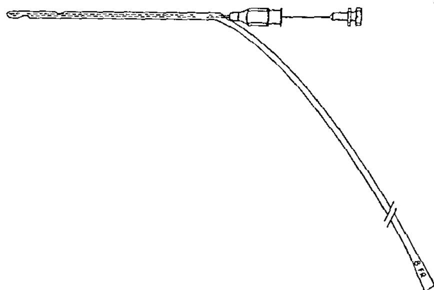


FIGURE 9. Technique of inserting a No. 20 spinal needle through a No. 8 French catheter in order to stiffen the catheter sufficiently for insertion into lateral ventricle (see text).

Ingraham and Matson should be routine. Care should be taken to obtain a perfectly straight antero-posterior view showing the fourth ventricle, it may show a lateral shift or the air may actually outline a tumor projecting down into the ventricle. One must be careful not to mistake the choroid plexus which hangs down into the fourth ventricle for a neoplasm in that region. Sometimes the dilated aqueduct is displaced upward or forward by a tumor in the posterior fossa. Similarly there may be elevations of the tentorium with displacement of usually one and occasionally both occipital horns of the lateral ventricles. This is seen in the presence of extremely large tumors usually astrocytomas in one cerebellar hemisphere. If the amount of air entering the ventricle is inadequate to permit localization of the tumor the ventriculostomy tube may be left in place and the x-ray studies repeated twenty-four hours later. In this interim sufficient air may enter the system Occasionally after performing a ventriculostomy and ventriculographic studies more information is necessary; it is possible to combine the ventriculogram with a pneumoencephalogram. The ventric-

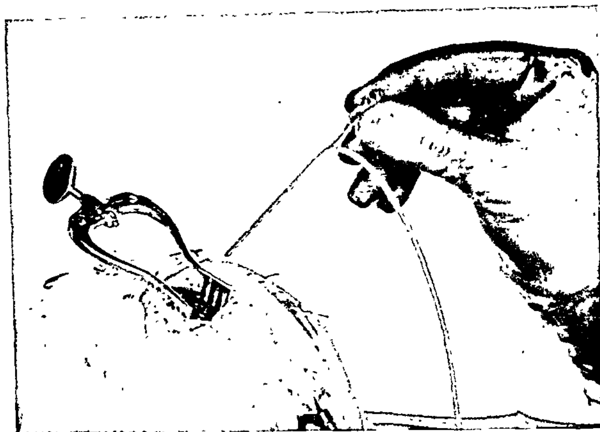


FIGURE 10. Method of inserting catheter into ventricle for drainage (see text).

ulostomy tube acts as a safeguarding decompression valve. Under these circumstances the dangers of pneumoencephalography in patients with brain tumors are minimal. But to emphasize a point, pneumoencephalography alone is done only in cases of suspected cerebellopontine angle tumors or in brain stem tumors in which there is no evidence of increased intracranial pressure. In these cases the cisterna around the pons, the aqueduct, the fourth ventricle and the cisterna magna can be visualized better than in ventriculography. However, if there is any doubt that increased intracranial pressure exists, even in these instances, pneumoencephalography is preceded by ventriculostomy. Angiography in this clinic is carried out if there is definite lateralization of the lesion, if it is suspected that the patient might have a vascular malformation, or if in the differential diagnosis the possibility of an intracranial abscess is entertained. In the latter instance ventriculography and of course pneumoencephalography is avoided so that the risk of any possible rupture of the abscess by change in intracerebral dynamics is obviated.

Anesthesia

After the lesion is localized plans must be made for its removal. Prior to initiating anesthesia certain precautions should be taken. The determination of blood counts, urinalysis and chest roentgenograms should be routine in

carina so that only one lung is aerated. The aeration of both lungs must be checked after positioning of the child irrespective of whether he is turned over in the face down position, in a supine position, or if he is placed in a sitting up position.

Supratentorial Tumors

Tumors arising in the supratentorial region are divided into four groups: (1) tumors in the cerebral hemisphere; (2) tumors in the hypophyseal region; (3) tumors in the pineal region, and (4) a miscellaneous group of tumors. Obviously this division is made only for purposes of easier discussion and not because the lesions in any one area comprise an entirely distinct entity, although pineal and hypophyseal tumors come close to it. There is considerable overlap from one area to the next in growth characteristics, clinical syndromes and in histological types.

Tumors in the Cerebral Hemispheres

Tumors arising within the cerebral hemispheres in children comprise about 25% to 30% of all intracranial tumors. They are predominantly gliomas, in fact about 85% of them are gliomas, with the other 15% being comprised of tumors of various histological types (angioblastomas, granulomas, dermoids, fibroblastomas, and metastatic tumors). The age distribution of the gliomas is shown in Figure 11. The reason for the greater number occurring in the two to three year age group is that most of the patients with ependymomas were of this age. Perhaps with a larger series this would have levelled off and the relative frequency would have been about equal during each year.

The relative frequency of the various histological types of gliomas occurring in the supratentorial region is given in Figure 12. There are obvious



FIGURE 11. Age distribution of supratentorial gliomas.

Histological Type	*Number*	*Total*
Astrocytoma	25	31.2%
Astroblastoma	6	7.5%
Glioblastoma multiforme	14	17.5%
Oligodendroglioma	5	6.2%
Spongioblastoma polare	4	5. %
Ependymoma	8	10. %
Ependymoblastoma	8	10. %
Pinealoma	10	12.5%
	80	100 %

FIGURE 12. Supratentorial gliomas—relative frequency according to histological diagnosis.

differences in frequency of the various types in children compared to adults; glioblastomas are comparatively rare cerebral tumors in children and pinealomas are fairly frequent.

Symptomatology

The symptoms and signs produced by a glioma in the cerebrum are dependent upon the exact location of the lesion, but usually they fall into the two general categories: (1) those of increased intracranial pressure, and (2) those of local involvement of the cerebrum. The tumors located more peripherally usually do not block the flow of cerebrospinal fluid so the initial symptoms seldom are those of increased pressure. Rather the initial symptoms are those of a change in the child's personality. The onset of this is usually so insidious that close scrutiny is necessary and every minor complaint offered by the child or suggested by the mother must be carefully weighed. Many times, and these changes unfortunately become apparent only by hind site, the child becomes listless, cares to play less enthusiastically, becomes withdrawn, or occasionally more irritable and difficult to manage, all changes that one is apt to attribute to psychological causes. In dominant hemispheral lesions, a change in handedness, hesitancy in speech, or difficulty in reading may occur. A weakness of an arm may manifest itself by improper table manners or if the child compensates well, the handling of the fork or spoon may be changed to the opposite hand. The child may limp, but a more frequent observation is that he stumbles a bit, runs less well, tends to walk less due to easy fatigability. It is interesting that headache is a relatively uncommon (20% to 30%) initial symptom, but occurs sometimes during the illness in most children (80% to 90%) This is in contrast to vomiting, which seldom occurs even late in the course of events Convulsive seizures are frequent; they may occur with lesions located anywhere in the cerebrum; they may be generalized or focal Attacks of petit-mal have been observed with

frontal and with temporal lobe tumors. Since seizures may occur in children from a large variety of causes they are of less diagnostic value than in adults, but a seizure occurring at any age warrants consideration and should not be casually brushed off as not pertinent. Because of the peculiarities of symptoms and signs in children compared to adults, it is believed a more liberal attitude toward supplementary diagnostic procedures is indicated in youngsters.

Papilledema should be looked for—the idea that it does not occur due to the tendency for the suture lines to widen or to the greater compressability of a child's brain is erroneous. It was present in 40% of the patients in this series with temporal lobe tumors, in 66% of those with frontal lobe tumors, 70% of parietal and 100% of those with occipital lobe tumors. Other signs include reflex changes, astereognosis, visual field defects and, of course, the well recognized changes of enlarging head circumference and bulging anterior fontanelle most often observed in infants. Supplementary diagnostic procedures invariably are necessary to pin down the diagnosis and to establish localization. These include routine blood and urine studies, skull roentgenograms, electroencephalography, angiography, and/or ventriculography. It is emphasized that angiography may give the surgeon the advantage of knowledge concerning the location of the vascular supply to the tumor and that with this knowledge a more complete removal of the tumor may be possible. It is also emphasized that the carotid system can be filled out quite readily and regularly with the reflux brachial angiographic technique especially in children under six to eight years of age.

Surgical Considerations

After it is decided a cerebral lesion is present, the technique of operation is essentially the same as that used for adults with similar lesions. The procedures are done with the patient in a supine position unless the occipital region is to be approached in which even a lateral decubitus position is used. The face down (prone) position need seldom if ever be used for a cerebral lesion. Anesthesia generally used is pentothal plus relaxant agent plus supplementary nitrous oxide. The skin is infiltrated with 1% procaine or xylocaine containing 5 to 6 drops per ounce of epinephrine. After the bone flap is turned the dura is inspected and palpated for evidence of tumor. If tumor is evident, the overlying dura is opened, but more frequently there is no evidence of a lesion that is discernable through the dura. To open the dura widely in the face of increased intracranial pressure invites disaster in all hands except those of the most experienced and fortunate surgeons. For subcortical lesions a far better technique is to make a small 2 x 2 mm. opening in the dura, locating this opening immediately over the presumed tumor. A ventricular needle is inserted and a

biopsy taken. Evidence of neoplasm, adjacent edematous tissue, or normal brain tumor can be had by inspection of the biopsy material. If in doubt of the nature of the biopsy a rapid frozen section can be made. In this clinic the technique of fluorescing biopsies of the suspected tissue with an ultraviolet light after injection of sodium fluorescein is routine and is believed more sensitive for identifying edematous or neoplastic tissue than histological examination. The most effectual and efficient source of ultraviolet light is a CH-4 mercury spot light with a Wood's filter. Several commercial models are available, the model used in this clinic has the lamp separated by a long cord from the transformer unit. This makes it possible to bring the shielded lamp up to the operating field without transporting the heavy transformer. It is the policy to inject intravenously from 3 to 5 cc. of 20% sodium fluorescein after the patient is asleep and just prior to making the skin incision. The amount injected depends upon the size of the patient. If the tumor is not located on the first biopsy, the procedure is repeated with insertion of the needle into the next most likely site. After the lesion is located, and if the pressure is such that wide opening of the overlying dura is hazardous (and the most likely hazard is bulging and possible rupture of the cortex), an internal decompression of the tumor is done. Perhaps a cyst can be aspirated with the needle, if not, the dura is opened a bit more, maybe 3 by 2 cm., sufficient only to enable one to insert the suction tip and remove the cortex overlying the tumor. The tumor is then entered and an internal decompression accomplished. As the increased intracranial pressure declines, the dura can be opened widely permitting a more extensive removal of the lesion preferably by lobectomy technique.

Of course, lobectomy is applicable only to frontal, temporal, and occipital lobe lesions, and unfortunately, not always with these. If the lesion is in the dominant hemisphere and in areas highly important for reasonable functioning of the child, care is taken not to go appreciably beyond the grossly visible confines of the tumor. An exception to this rule is the unusual case in which it is believed a cure can be accomplished by more radical resection. Children recover from neurologic dysfunction to an extent far exceeding the adult. It is believed the remaining cerebral tissue in children will quite readily assume the functions usually carried out by the excised tissue. Nielson has alluded to this assumption of function by his statement that aphasia does not develop in infants from a unilateral lesion regardless of laterality. This ability to recover function seems to hold true throughout the first decade at least. In the final analysis it is the judgement of the surgeon at the time of operation that dictates the type and the extent of the resection. The factors important in this decision include the preoperative condition of the patient, tolerance of the patient

to surgery, apparent histological type of the lesion, the location, and the extent of the lesion. If the patient's preoperative condition is very poor and the child does not seem to be tolerating well the turning of a bone flap, it is advisable to do the operative procedure that is least traumatic but still consistent with adequate reduction of the intracranial pressure. If a large cyst is encountered it should be tapped and the final or definitive surgery done some days or weeks later. It is imperative, however, always *to obtain sufficient decompression to permit a normal flow* of cerebrospinal fluid. It is believed the concept of "biopsy of the lesion and if it proves to be a glioma to close the incision" should be condemned.

Gliomas vary in growth characteristics and location according to histological type. Obviously the surgical approach is dependent a great deal on the histology. For this reason the various tumor types will be discussed separately.

Astrocytomas

Ford stated that 40% of all gliomas in children are astrocytomas and Cushing (1932) found 35% of 116 gliomas in children to be astrocytomas. Contrary to their location in adults, in children astrocytomas are located predominantly in the cerebellum (40%) or brain stem (10%) and only 30% in the cerebrum. There is no sex predilection; there is no age predilection for these tumors.

Upon their gross appearance, astrocytomas are divided into two types: *The cystic type* appears as a large smooth walled cavity filled with yellow fluid. The wall is made of compressed brain tissue except in one area where there is a firm well circumscribed nodule of tumor tissue (mural nodule). This was known as the gliomatous cyst to the earlier investigators. The cystic fluid is transudate; it coagulates on standing. The *noncystic type* (cystic glioma of the earlier investigators) is a diffuse, infiltrative lesion that may contain multiple small cysts but these cysts are entirely surrounded by tumor tissue. Thusly differentiated about 40% of all astrocytomas in children are of the cystic and 60% are of the noncystic type. The cystic astrocytomas occur only in the cerebellum, the noncystic occur with about equal frequency both above and below the tentorium. Both types of astrocytomas are slowly growing and difficult to differentiate histologically. It is the gross appearance upon which the above classification is made. Calcareous deposits of size sufficient to be seen in roentgenograms may occasionally be found in astrocytomas of either the cystic or noncystic type.

Astrocytomas are tumors composed predominantly of cells of adult structure—the astrocyte. On the basis of the histological appearance Penfield (1931) divided the astrocytomas into three types: (1) *pilocytic;* (2) *gemistocytic;* and (3) *diffusum.* He believes the three types, in addition

to differences in histological appearance, show clinical differentiation although most observers do not agree with this (Bailey 1932, Bucy, Baker). The pilocytic type is composed of small elongated cells, is firm and fairly well circumscribed, is slowly growing, occurs both in the cerebellum and cerebrum and when located in the cerebellum is the type that forms the cystic astrocytoma. The gemistocytic type is more rapidly growing, contains multiple small cysts, is found only in the cerebrum and is composed of more irregular but small cells with fairly deeply staining nuclei. The astrocytoma diffusum is composed of small cells with small rounded nuclei, has an indefinite tumor boundary, is seldom cystic, and found only in the cerebrum. Bailey and Cushing have differentiated two histological types of astrocytomas, the protoplasmic and the fibrillary, but they state there are many gradations between them. The protoplasmic tend to be located near the surface whereas the fibrillary are in the deeper white matter and in the region of the basal ganglia, the third ventricle, and the brain stem. These fibrillary astrocytomas are comparable to the pilocytic and diffusum type of Penfield whereas the protoplasmic resemble more the

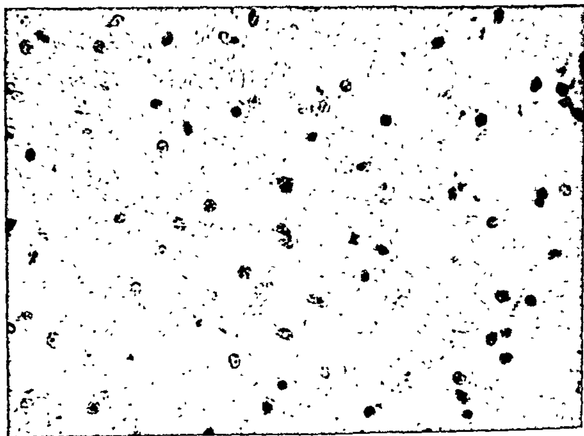


FIGURE 13. Photomicrograph of a fibrillary type astrocytoma. Note the lack of background staining. The cells are regular, consistent appearing, and in this field not very numerous.



FIGURE 14. Astrocytoma left basalar ganglia region. The tumor spreads diffusely into the adjacent brain tissue; a definite boundary cannot be distinguished. There is a shift of the ventricular system to the right due to the mass of the neoplasm and the surrounding edema.

gemistocytic type of Penfield. There is a strong resemblance between the fibrillary (pilocytic) astrocytomas and the polar spongioblastomas.

In this series of seventy-three astrocytomas, twenty-five were located in the supratentorial region. Seven were predominantly or totally in the frontal, three in the parietal, two in the occipital, and five in the temporal regions. Three arose in the suprasellar region (optic nerve gliomas) and five were located in and around the third ventricle. All were of the non-cystic or diffuse type. The average age at the time of operation was 7.2 years. The average duration of symptoms before hospital admission for the hemispheral or peripheral lesions was seven months and for those located closer to the third ventricle was 10.6 months.

Astrocytomas located in the peripheral part of the cerebral hemispheres usually appear fairly well circumscribed although they may blend with and infiltrate the adjacent brain tissue so that a boundary between the normal and the tumor tissue cannot be determined. They are fairly slowly growing and temporarily amenable to surgery. However, all those in this series were so infiltrative that permanent cure was out of the question. It is believed that lobectomy as the primary procedure might afford a cure

in some patients. The tumors are moderately responsive to deep roentgen therapy and following surgery a course of deep therapy should be considered.

The postoperative survival time is extremely variable but generally a period of two to three years prior to recurrence can be expected and often times this extends to five to ten years. This interval is dependent upon the location of the lesion. Those located in the frontal area can be treated with lobectomy plus roentgen therapy and longer survival is expected than for those located in the parietal or temporal lobes where the extent of decompression is necessarily less. When recurrence does become manifest a second operative procedure generally is advised and this can again be followed by deep roentgen therapy. If the astrocytoma is deep, in the region of the basal ganglia, a frequent symptom is hemiparesis or paralysis and whereas this may clear up following subtotal removal of the neoplasm, only a very temporary (one to three year) period of relief is anticipated. Second operative procedures to lesions in this area are never advised due to the very high morbidity expected. A course of deep roentgen therapy may afford some temporary relief.

Astroblastomas

Astroblastomas are quite rare tumors that occur most frequently in the white centrum of the cerebral hemispheres although cerebellar and optic nerve astroblastomas have been reported. Only to those tumors in which the astroblast definitely is the predominant cell is the term astroblastoma applied. According to Bailey 1932, this tumor type stands midway between the astrocytoma and the glioblastoma.

On gross examination astroblastomas appear as huge poorly circumscribed and infiltrative lesions. They may be cystic and contain soft gelatinous areas and areas of hemorrhage. On histological examination, astroblastomas are difficult to differentiate from glioblastomas. The cells vary greatly in size and shape but usually are triangular and each cell possesses a sucker foot that is attached to a blood vessel. With routine hematoxylin and eosin the cytoplasm does not stain and the nuclei of the astroblasts appear therefore to radiate out from the blood vessels in the form of pseudorosettes. Astrocytes and spongioblasts may be present. Not infrequently astroblastomas are very vascular and the vessels may show some severe proliferative changes, especially in the adventitia.

Astroblastomas are fairly rapidly growing, malignant infiltrative lesions. The duration of preoperative symptoms in this series of six cases was only six months with a range from three weeks to two years. All patients had symptoms of general malaise, easy fatigability, weakness and listlessness. This group of symptoms seemed more frequent in patients with astro-

blastomas than any other tumor type other than the glioblastomas. In one of the tumors there was sufficient calcium to be visible in roentgenograms, but this is an extremely rare occurrence. Like the glioblastomas, the astroblastomas are invasive diffuse tumors that infiltrate along fiber tracts. The accepted method of treatment is surgical removal of as much of the tumor as possible but complete removal could not be obtained in any of the patients in this series. Roentgen irradiation following surgery seemed to have limited if any effect. The average postoperative duration of life is about one to two years.

Glioblastoma Multiforme

In this series there were twenty-six patients with tumors diagnosed as glioblastoma multiforme. In twelve patients the tumor was located in the brain stem and in fourteen the tumor was in the cerebrum. The favorite cerebral location was the temporal lobe but these tumors are so invasive that almost always more than one lobe was involved. Glioblastomas usually arise in the cerebral white matter but rapidly grow out into cortex and



FIGURE 15. Photomicrograph of a glioblastoma. There are numerous gemistocytic cells, bizarre appearing cells, pleomorphism and alterations in appearance of vascular structures which are typical of this type tumor.

may even pass through the pia mater to form attachments to the dura. They tend to advance along nerve tracts down into the peduncle or cross via the corpus callosum to the contralateral hemisphere (Maxwell). On gross examination they appear as large multicolored, soft, necrotic, hemorrhagic infiltrative lesions. They have all the characteristics of uncontrolled malignancy. In adults they are usually solitary lesions but in children they frequently are multiple (Elsberg and Globus, Irish, Courville 1937).

Histologically these tumors are characterized by a multiform appearance of the cells and of the vascular pattern. The cells vary from small, rather mature astrocytes to large multinucleated giant cells. The predominant cell type is a spindle-shaped spongioblast, but astrocytes, astroblasts, medulloblasts, amoeboid, and giant cells many of which may be multinucleated and often of a bizarre shape are usually present. Mitoses are numerous. The blood vessels show characteristic changes such as dilatation, tortuosity, hyalinization, and proliferation of intima and adventitia. The lumen of the blood vessels may be invaded by tumor cells resulting in hemorrhages and thromboses. Degenerative areas and necrotic areas are common. A pallisading of cells occurs along these necrotic areas and this is one of the outstanding features of glioblastomas.

There are no signs or symptoms typical of a glioblastoma except a rather



FIGURE 16. Gross appearance of a glioblastoma located primarily in the left temporal lobe but also extending into the adjacent lobes. This has more of a hemorrhagic appearance than the usual glioblastoma.

rapid clinical course of about two to four months' duration. Since the most frequent location in children is in the brain stem, the most common early symptoms are cranial nerve palsies, frequently multiple, and only late may signs of aqueduct obstruction be noted. There may be direct invasion of the respiratory, cardiac, or vomiting centers by tumor tissue. In addition, signs of easy fatigue, slight lethargy, and mental and emotional changes occur. When the lesions occur out in the cerebrum there is rapid development of evidence of loss of neurological function (motor, sensory, mentation, seizures) and increased intracranial pressure.

It is felt that radical surgical excision of all recognizable tumor tissue is the best method of therapy for those located out in the cerebrum. It is believed that extremely large areas of decompression of the brain should be obtained. Deep roentgen therapy has been advised by some, but in this clinic the response to radiation therapy has been insufficient to warrant its routine use. Irrespective of the method of treatment, the prognosis is poor; postoperative life expectancy is less than a year.

Spongioblastoma Polare

The term spongioblastoma polare is given to those tumors of a transitional nature that are comprised of uni- and bipolar cells arranged in sheaths or bundles and running in parallel rows. The cells are mature. The nuclei contain numerous coarse masses of chromatin. The cytoplasm is not abundant and the cells are pyriform or spindle shaped. Occasionally astrocytes may be seen in these tumors. On gross examination spongioblastomas are difficult to differentiate from normal brain tissue. This is especially true when they are located along the brain stem. They are firm, rather pale gray to yellow in color, very poorly defined lesions. The actual size of the lesion is difficult to determine because of the infiltrative character. Sometimes these tumors are difficult to distinguish from pilocytic astrocytomas because both tumors have similar elongated cells. This is especially true when the astrocytoma is located in the region of the basal ganglia or brain stem. It has been postulated that this elongated type astrocyte is so shaped because of compression by fiber tracts. It has been observed that cells of the spongioblastoma polare growing in tissue culture appear similar to astrocytes. There is no doubt that the two types of tumors are very similar and may even be the same.

Spongioblastomata polare are slowly growing. They tend to infiltrate along fiber bundles and this tendency along with their frequently inaccessible location often makes surgical excision impossible. There are no specific diagnostic features except that they cause symptoms of long duration with involvement of the optic chiasm, basal ganglia, or brain stem. There frequently is early involvement of the cranial nerves and only late in the course

do symptoms of increased intracranial pressure become manifest. This sequence of symptoms is of special diagnostic value. Spongioblastoma polare may be associated with systemic evidence of von Recklinghausen's disease. They often contain sufficient calcium to be visible in roentgenograms (Bailey and Eisenhardt).

Spongioblastomata polare are infrequently encountered tumors, only five being present in this series of 195 gliomas in children. One of the tumors was located in the pons, one in the optic nerve chiasm, and three in the third ventricular region. Others have reported a similar distribution, i.e., Bailey (1927) stated that they are predominant in the region of the third ventricle and optic chiasm and Hamby reported a calcified spongioblastoma polare filling most of the third ventricle. The age of occurrence of spongioblastoma polare varies but the majority are in children. Pilcher reported

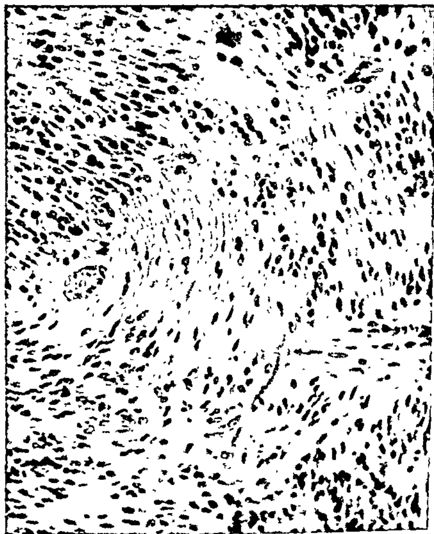


FIGURE 17. Photomicrograph of a spongioblastoma polare. The characteristic uni- and bipolar cells arranged in sheaths or bundles and running in parallel rows are evident. When the lesion is less cellular these tumors are difficult to differentiate from astrocytomas and occasionally from normal brain tissue.

eight of eleven to be in children under sixteen years of age with an age range from three to thirty-five years. There is no sex predilection.

These tumors should be removed as completely as possible if they are at all accessible. They are slowly growing and in case a cure cannot be obtained with excision a long survival can be anticipated. Those located in the brain stem are, of course, inaccessible. Deep roentgen radiation is not of much value.

Ependymomas

Ependymomas in children are generally considered posterior fossa lesions but actually they arise from the ependyma and subependymal tissue anywhere along the ventricular system. Of thirty-two ependymomas in this series, nine were in the lateral ventricular region and fifteen were in the fourth ventricular area. The stem cell of these tumors is the ependymal cell and the histological appearance is dependent upon the stage of differentiation of the stem ependymal spongioblast. On the basis of a histological and clinical differentiation Kernohan and Fletcher-Kernohan have divided the ependymomas into four types (1) *Papillomas of the choroid plexus;* (2) *myxopapillary ependymomas;* (3) *epithelial ependymomas;* and (4) *cellular ependymomas.* More recently they have added another type, *ependymoblastoma,* a type previously dropped by Bailey 1932.

Papillomas of the Choroid Plexus

The papillomas of the choroid plexus simulate in appearance the normal plexus. They are papillomatous growths composed of a central core of loose connective tissue and small blood vessels surrounded by a layer of high columnar cells that contain mucin. Grossly they are irregular, red to brown in color, well circumscribed lesions lying entirely within the ventricle as shown in Figure 19.

Papillomas seem to arise either in the glomus of the plexus in the lateral ventricle or from the plexus along the anterior medullar velum of the fourth ventricle. They do not invade the adjacent cerebral or cerebellar tissue but gradually enlarge, eventually entirely filling the ventricle and even crossing from one lateral ventricle through the third ventricle into the contralateral ventricle. However, it is much more usual that symptoms of increased intracranial pressure develop prior to such extensive growth. The increased pressure is due to block of flow of cerebrospinal fluid although Kahn and also Matson have reported a communicating type hydrocephalus with increased intracranial pressure from hypersecretion of a papilloma of the plexus. A positive diagnosis can only be made from diagnostic air studies, preferentially ventriculography. Pneumoencephalography can be especially hazardous in these children because the papillomatous mass can move forward to block off a ventricle when there is release

of cerebrospinal fluid pressure below the lesion. On ventriculography the papillomatous mass may stand out, well delineated in a ventricle filled with air.

The therapy is surgical resection which usually is possible when the lesion is in the lateral ventricle. One must be somewhat less optimistic for those encountered in the fourth ventricle. For those located in the lateral ventricle, the best site of approach is in the posterior parietal area about the junction of the parietal, occipital, and temporal lobes. It is recommended that a bone flap of at least 6 x 6 cm. in size be elevated. If there is greatly increased intracranial pressure it is probably wisest to make an opening in the dura large enough only to permit insertion of a ventricular needle through this opening into the involved ventricle. This will allow a decompression but only sufficient fluid to accomplish a decompression should be drawn off. If all the ventricular fluid is allowed to drain off, it may be very difficult to get back into the ventricle and also it makes the dissection of the tumor more difficult. An incision in the cortex following the needle tract is usually the best means of entrance into the ventricle. The walls of the enlarged ventricle should be supported by a malleable retractor.



FIGURE 18. Photomicrograph of a papilloma of the choroid plexus. This is histologically and grossly a benign lesion.



FIGURE 19. Gross specimen of papilloma of the choroid plexus of the left lateral ventricle. The lesion had not infiltrated into the ependyma or subependymal region and was surgically resectable.

If one can then reach the glomus area from where the major blood supply arises, the vessels can be clipped and the tumor mass may be reduced in size permitting good visualization throughout the ventricle. It is best if the tumor can be removed intact but piece-meal removal must sometimes be carried out. Care must be taken not to leave fragments of tumor tissue that might break off and drift down into the temporal horn or forward into the anterior horn. Following the removal of the tumor the ventricle should be washed out with saline solution and at the time of closure the ventricle should be left full of saline so that there will be no collapse of the cerebral mantel. An unusual postoperative complication is that observed whenever intraventricular surgery is done in a child with hydrocephalus; that is, the cerebral mantel may collapse tearing the veins leading from the cortex to the dura with the development of a subdural hematoma or subdural hygroma. This is mentioned just as a word of caution. Occasionally these tumors are myxopapillomatous and invasion of the adjacent cerebral tissue may occur. The prognosis is good unless there is invasion into the adjacent cerebral tissue which, in these tumors, infers greater malignancy and total removal may not be possible. In this event postoperative deep

roentgen therapy is advisable. The same is true if the lesion proves to be an adenocarcinoma. Although this latter histological type seems rather indefinite and none have been encountered in this clinic, Ingraham and Matson list six of their nine tumors of the choroid plexus as adenocarcinomas. In the true papilloma choroidium a cure can usually be accomplished by surgical excision.

Myxopapillary Ependymomas

Myxopapillary ependymomas are similar to papillomas of the choroid plexus except the stroma of the papillary core undergoes some myxomatous degeneration and the surrounding layer of cells are cuboid and seldom contain mucin. According to histogenetic relationships these tumors are slowly growing lesions located in fairly accessable areas. However, none of the five tumors in this series were removed totally. They were unusually invasive of adjacent cerebral tissue, seemed to grow along the ventricular wall in the subependymal layer, and were entirely too extensive to permit total surgical removal. Likewise deep roentgen therapy seemed to be of very little avail. The longest survival period was two years and five months.

Epithelial Ependymomas

Epithelial ependymomas are composed of cuboidal cells, many of which are arranged in canals resembling the primitive central canal of the spinal cord. The canals are empty and so differ from the myxopapillary type. The cells may grow side by side in a pavement-like pattern which resembles epithelium and it is because of this arrangement that the term epithelial is given. Grossly these tumors may appear well circumscribed but invariably they infiltrate out into the cerebral tissue so that total removal is seldom if ever possible. They are red to gray in color, rather vascular, may contain small cysts although usually they are fairly firm tumors. When located in the posterior fossa they arise from the ependyma of the floor of the fourth ventricle, extend out into the ventricle, and do not invade the cerebellar hemisphere or vermis. Total removal is not possible because the infiltrative area is down into the pons and medulla. Total removal can only be accomplished with destruction of vital medullary centers.

Cellular Ependymomas

Cellular ependymomas are composed of closely packed cells with large oval nuclei and abundant cytoplasm. Mitoses are fairly common and the differentiation between these and the ependymoblastomas is difficult. Both contain degenerative cystic as well as hemorrhagic and necrotic areas. In the ependymoblastoma mitoses are more frequent. There is not a great deal of difference clinically between these lesions and the glioblastomas.

The vascular changes typical of glioblastomas do not occur in cellular ependymomas and ependymoblastomas, but all three are highly invasive, rapidly growing malignant tumors. When located in the cerebral hemispheres at least an extensive decompression of the tumor should be accomplished and if the tumor is so located a lobectomy should be done. The general prognosis is poor; total removal probably has never been ac-



FIGURE 20. Photomicrographs of a myxopapillary ependymoma (a), and an epithelial ependymoma (b). a. Myxopapillary ependymoma. These are similar to the papillomas of the choroid plexus with the exception that the papillary core undergoes some myxomatous degeneration. They are histologically benign but there usually is sufficient involvement of the subependymal region to preclude total surgical removal. b. Epithelial ependymoma. Note the gliovascular pattern characteristic of these tumors. They are infiltrative, fairly malignant lesions that seldom, if ever, can be totally removed by surgery.

complished. Postoperative length of life is one to two years at the most. Deep roentgen therapy is of questionable value but as in most all ependymomas except papillomas it is given with hopeful heart.

Oligodendrogliomas

In this series of 195 gliomas in children, only five were oligodendrogliomas. All were located supratentorially in the frontal, parietal, or occipital regions. Whereas oligodendrogliomas occur predominantly in adults they have been found in very young children. Hughes and Couper reported an oligodendroglioma in an eight month old child who had symptoms dating

back to the age of two weeks. Critchley in 1925 collected from the literature 125 children with oligodendrogliomas. He found one in a child less than twelve months old. These tumors are composed of cells having a small rounded nucleus surrounded by a halo of cytoplasm. The cells may be divided into rows or cords but usually a pavement-block like appearance is presented. Astrocytes may be scattered throughout the tumor. On gross examination oligodendrogliomas are fairly firm, solid tumors in which calcium may be present. They appear rather circumscribed although not discretely encapsulated and in areas infiltrate into the surrounding brain tissue. Not infrequently they can be observed to infiltrate along in the subependymal region around the ventricle.

At the time of surgery, the tumors are found subcortically beneath a flattened and pale convolution. The line of cleavage between normal and tumor tissue is difficult to establish and total removal is seldom accomplished. The most propitious type of excision, if possible, is lobectomy and even if this is accomplished, recurrence is the rule because the tumor has infiltrated out along the ventricle. When oligodendrogliomas reach the ventricle, they often grow along the walls eventually reaching back as far as the aqueduct and fourth ventricle (Greenfield and Robertson). However, the prognosis is good for immediate relief of symptoms. These tumors usually are located in accessible areas of the brain, are slowly growing and even if not totally removed, relief of considerable duration is obtained. Some reports suggest these tumors are not as benign and slowly growing as is generally assumed. There is some evidence that suggests that the speed of growth increases late in their evolution and that operation serves to increase this rate of growth (Elvidge *et al.*).

In none of the children operated upon in this clinic has total excision of an oligodendroglioma been obtained, but total removal has been accomplished in a seventeen-year-old boy. In all of the children the tumor recurred. In two patients second and third attempts at total removal have been carried out but at each succeeding procedure it was apparent all the tumor could not be removed. However, with this method of re-operation, survivals of five to eight years duration were accomplished.

Neuroepithelioma and Medulloepithelioma

The neuroepithelioma and medulloepithelioma are considered together because of their relatively similar histological and clinical character. These tumors are composed of primitive spongioblasts characteristically arranged in rosettes about central canals. They are extremely rare tumors—Bucy and Muncie reported the case of a neuroepithelioma occurring in the cerebellum of a five-year-old boy and Bailey *et al.* (1939) reported one in the cerebellar region in a ten-month-old infant. There were none in the series of brain tumors seen in this clinic.

There is no characteristic symptomatology except that these tumors are probably the most malignant or rapidly growing of all gliomas. The pre-operative duration of symptoms ranges around four to six months. It is believed an attempt should be made to remove the lesion totally. At the time of surgery special care must be taken to prevent the rather friable lesion breaking off and spreading in the subarachnoid space. However, this is seldom prevented because the tumors invariably recur both at the original site and in implanted areas throughout the neural axis. Deep roentgen therapy seems to give some temporary relief but the general prognosis is extremely poor with very few of the patients surviving more than six to eight months.

Ganglioneuromas

The ganglioneuromas are extremely rare lesions. When they occur they have a predilection for the region of the third ventricle. However, not infrequently they are also found out in the centrum of the temporal lobe. Zülch (1938) reported five, all located in the cerebrum, among his series of 263 brain tumors in children. None were encountered in the University of Minnesota series. Kuhlenbeck and Haymaker have given an excellent review of these tumors.

Schmincke was the first to give an adequate description of these tumors in the central nervous system. In addition to the glial structure, the ganglioneuromas contain adult neurocytes which vary in size. The tumor cells can be identified by the large vesicular nuclei containing one or more large nucleoli with a prominant nuclear membrane and abundant cytoplasm that often appears hyalinized. The neurocytes are found in scattered groups or nests throughout the tumor. Probably better than the term ganglioneuroma is the one given to these tumors by Kernohan *et al.* in 1932 who suggested the term gangliocytoma because these tumors are composed predominantly of adult nerve cells.

Ganglioneuromas are usually slowly growing and present no typical diagnostic clinical signs or symptoms. If uncovered at surgery, and if they are accessible, an attempt should be made to excise them. Their general gross appearance varies considerably but usually they are fairly firm, gray to white lesions that seem to infiltrate the adjacent tissue. However, even though they do infiltrate out into the adjacent tissue the total area of involvement is not great and occasionally they can be totally removed. Radium or deep roentgen therapy is probably advisable in the irremoval cases. Total removal was not possible (Ford).

Tumors in the Hypophyseal Region

The predominant tumor occurring in the hypophyseal region in children is the craniopharyngioma, less common are gliomas of the optic nerve or

chiasm, parasellar meningiomas, gliomas arising in the frontal lobe and extending down into the suprasellar region, mixed tissue tumors, pituitary adenomas, and an occasional paraphyseal cyst which really is an anterior third ventricle lesion. All but the craniopharyngiomas are extremely rare. These tumors are grouped together because of the relatively similar clinical picture they present. The surgical approach to them varies depending upon the origin and extent of the lesion as well as upon the histological type.

Craniopharyngiomas

These tumors may become symptomatic at any age of life but the majority do so within the first two decades. In a series of twenty-one craniopharyngiomas reported by Beckman and Kubie, six (29%) were under sixteen years of age. Cushing (1932) reported twelve (34%) of thirty-five cases to be under fifteen years of age and in a series of 138 collected cases of Bailey, Buchanan and Bucy, sixty-six (48%) were under twenty years of age. Others have found similar age frequency (Frazier and Alpers 1931, Critchley and Ironsides, Ingraham and Matson). There is no specific sex predilection.

There have been several different names applied to these tumors (craniopharyngioma, Rathke pouch cyst, adamantinoma, and hypophyseal duct cyst). The term "craniopharyngioma" was introduced by Cushing and it is the one generally used since it is noncomittal of histological appearance but does designate the site or origin (Frazier and Alpers). Craniopharyngiomas arise from squamous cell rests that have their embryonic origin in the buccal (pharyngeal) epithelium and as a result of upward migration and rotation eventually are located in the superior anterior region of the anterior lobe of the hypophysis. These cell rests may proliferate and develop into a tumor that may be comprised of solid masses of cells or they may become cystic. According to the histological appearance Bailey (1932) divided these tumors into three types: (1) *Rathke pouch cyst;* (2) simple *squamous epithelioma,* and (3) *adamantinoma* or *ameloblastoma.* The Rathke pouch cysts are thin-walled cystic lesions in which the wall is composed of fibrous tissue containing areas of calcification and is lined by tall, ciliated, columnar epithelium. The cysts are filled with a clear fluid. They are usually limited to the pituitary fossa or adjacent areas. The simple squamous epitheliomas are cystic tumors whose walls are composed of fibrous tissue lined by stratified squamous epithelium grown into numerous papillomatous masses. The adamantinomas are larger, cystic or solid tumors varying from 2 to 8 cm. in size in which gross calcareous nodules may be seen. These tumors are composed of masses and columns of epithelial cells that resemble the embryonic enamel organ. They are large, adherent to adjacent structures and not removable surgically.



FIGURE 21. *(Top)* Photomicrograph of a craniopharyngioma (adamantinoma) with characteristic masses of columnar and stellate cells.

FIGURE 22. *(Bottom)* Craniopharyngioma extending up into the hypothalamus and even over into the right temporal lobe. This was obviously an inoperable tumor.

Symptoms produced by these lesions are due to the mass of the tumor producing increased intracranial pressure, to traction on pain sensitive structures or to local pressure on structures adjacent to the tumor (chiasm, hypothalamus, frontal lobe). The most frequent initial symptoms is headache. It is often associated with vomiting. The headache is diffuse and nonlocalized. The headache may be relieved by the child assuming the knee-chest position (Figure 4). Presumably the tumor in this instance is shifted forward by this maneuver, thereby relieving an obstruction in the third ventricle to the flow of cerebrospinal fluid. Frazier and Alpers reported the relative frequency of the first symptom in thirteen cases as follows:

Symptoms	*Number of Cases*
Headache	6
Arrested growth	3
Headache and decreased vision	1
Headache and somnolence	1
Decreased vision	1
Convulsive seizures	1

Evidence of endocrine dysfunction is frequently found and is of the *Fröhlich type. There is suppression of normal pituitary activity. Sexual* development is either slow or completely arrested. There is retarded growth (dwarfiism, smallness of bony structures, slowness of tooth development, etc.). The skin is soft, smooth, and often hairless. Neither acromegaly nor gigantism are seen in patients with these lesions (Witterman, Critchley and Ironsides).

Symptoms resulting from pressure on the hypothalamic region such as *somnolence, diabetes insipidus, decreased* glucose tolerance, and lowered basal metabolic rate, are frequent. According to Peet the somnolence is due to a subnormal metabolism rather than a direct involvement of any centers of consciousness by the tumor. As these tumors enlarge they exert pressure onto the optic chiasm or nerves. The usual visual field defect is a bitemperal hemianopsia but frequently the defect is irregular. The type of visual field defect depends upon the exact location and direction of extension of the tumor. Occasionally the field defect is restricted to one eye. *If there is direct pressure on an optic nerve, visual loss from primary* optic atrophy may occur. In Ingraham and Matson's series of twenty-one *patients, fourteen (66%)* demonstrated visual disturbance at the time of hospitalization.

Roentgenographic evidence of calcium in the suprasellar region is common in craniopharyngiomas. The calcium may be finely stippled and fairly well dispersed throughout the tumor or may be in heavy, coarse clumps. *Occasionally* the calcium is present only in the wall (Rathke pouch cyst) and appears as a curvilineal shadow in roentgenograms. MacKenzie and

Sosman made a correct diagnosis by radiological evidence of suprasellar calcification in 71% of their cases.

These tumors are histologically benign, but only a subtotal surgical removal can be accomplished because of their location and adherence to either the adjacent hypothalamic region or the carotid arteries. Recurrence of symptoms can be expected after a period of one to five years. Repeated surgical removal is possible but the ultimate prognosis for life is poor.

When a tumor is suspected in the suprasellar region, it is advisable to carry out ventriculography or carotid angiography rather than direct exposure of this area. The reason for the air study is to ascertain the extent of growth of the tumor. For example, it is advantageous to know if it is extending back into the third ventricle with the possibility of closing off the flow of fluid at the foramen of Monro or whether it is extending up into the frontal lobe with a projection into one or the other frontal horns. In this event, rather than using the classical transfrontal approach with elevation of the frontal lobe, it is probably wise to attack the lesion



FIGURE 23. Pneumoencephalogram of a patient with a craniopharyngioma showing the tumor mass projecting up into the anterior third ventricle region.

transventricularly. The purpose of doing carotid angiograms is to ascertain whether or not the neoplasm involves the arteries. Occasionally the arteries can be so displaced by the neoplasm that they are not recognized at surgery and can be injured.

For the usual tumor located in the suprasellar region, the classical approach to it is via a transfrontal craniotomy. Since inclusion of the incision within the hairline is most desirable, a coronal type skin incision is preferred. This type of incision should follow in a gental curve just below the hairline. Never should it be carried down into the temporal region in such a manner that any of the fibers of the facial nerve are severed. By the same token, it must be carried down sufficiently far to make unnecessary a stretching of the skin when it is turned down over the eyebrow region for this stretching might also injure the fibers of the facial nerve that innervate the frontalis muscle. To facilitate turning the skin flap the center part of the incision bends forward at the midline of the head. As in all scalp incisions in children, it is wise to infiltrate the galeal region with 1% procaine containing five to six drops of epinephrine per ounce. Raney clips are placed along the skin margins—no complications relative to ischemia of the skin have been observed, at least any due to the Raney clips, but necrosis may result from too enthusiastic coagulation of the vessels external to the galea. The bone flap is turned down in the usual manner. It should be placed so that the medial edge is along the midline and the anterior edge skirts the frontal sinus. In case the sinus is entered, a piece of muscle or gelfoam is placed over this opening. This is left in place permanently.

An extradural approach to the chiasmal region is made, the dura being incised along the sphenoid wing. The original opening in the dura is made out laterally along the sphenoid wing almost at the pterion. There are only two complicating features. There is a small vessel that invariably runs from the lateral portion of the roof of the orbit to the overlying dura. This can be closed by tamponade with soft bone wax or with the cutting current of the electrocoagulation unit. Also incision through the dura should be made as close as possible to the edge of the bone and care taken not to go too deeply, for underlying this are the vessels of the sylvian area. Occasionally a small vein extends from these sylvian vessels out to the dura and an incision made too far laterally may necessitate coagulation which in turn may lead to rupturing of more of the adjacent veins.

In case there is greatly increased intracranial pressure, making the elevation of the frontal lobe difficult, the pressure can be reduced in various ways. One method is to do a lumbar puncture. This, however, is seldom resorted to in this clinic. More frequently a small opening is made in the dura overlying the frontal lobe and cerebrospinal fluid permitted to drain

out through this opening. In case this is inadequate and it is believed the ventricle can be cannulated, this is then done. After the dura along the sphenoid wing is opened, cerebrospinal fluid will escape. By advancing slowly permitting cerebrospinal fluid to drain, always opening the dura a bit more, one can eventually gain all the room necessary in which to operate. The medial portion of this dural incision should be carried upward, back along the midline. This facilitates the exposure. After the suspected neoplasm has been visualized, the temptation is to incise the capsule with a knife and to suck out the contents. A wiser procedure is to insert the tip of a No. 22 spinal needle to which is attached a two to five cc. syringe. In case blood is encountered, the approach should be modified from that used if the typical oily fluid of the craniopharyngioma is obtained. Perhaps the lesion is an aneurysm rather than a craniopharyngioma (even though angiography has not shown an aneurysm), or the cyst sac has been stretched over a medially placed carotid artery. This needle technique is so easy and so assuring that it is considered foolish to dispense with it no matter how self-assured the surgeon is.

After the capsule of the tumor has been entered, care is taken not to spill any of the content for it can be very irritating if lost into the subarachnoid space. An aseptic chemical meningitis may be produced. Following the aspiration of the fluid content a currette is used along with the suction tip to remove the more firm and somewhat fixed caseous lining of the suprasellar cyst. The cyst wall can often be delivered partially by gentle tugging, but only after the cyst contents are removed. Obviously too much traction on the wall that is adherent to the hypothalamus may result in intractable damage. Another danger is overly zealous traction via the cyst wall onto the carotid arteries or the optic chiasm. Total removal of a craniopharyngioma is seldom accomplished—and only then when the cyst is small. Several methods have been tried to drain the cyst fluid either into the frontal sinus or into the subcutaneous tissue so that repeated aspiration of the fluid is possible. These procedures are done, of course, only when the content of the cyst is fluid and when the cyst is of sufficient size to make such an anastomosis possible.

In case the craniopharyngioma cyst is extremely large so that it projects up into the frontal lobe displacing the anterior horn of the lateral ventricle or even projecting into the ventricle itself it may be wisest to approach the tumor transventricularly rather than via the usual subfrontal approach. If the transventricular approach is used, the skin incision should be within the hairline, the bone flap should expose an area of the frontal lobe at least 5 x 7 cm. in size but the dural incision and the original incision through the cerebral mantle into the ventricle can be small. The dural incision should be about 5 to 6 cm. long and made in a semicircular shape. Following this

the ventricle should be located by insertion of a blunt needle (the baby type ventricular needle of Matson is excellent) into the anterior or tip portion of the ventricle. Very little or no cerebrospinal fluid should be permitted to escape for if it does, difficulty may be experienced in finding and entering the ventricle. The needle tract often can be followed down into the ventricle. Whether or not one takes a core of the cerebral mantel or makes a linear incision into it in order to enter the ventricle is immaterial. The incision, however, should go through the middle portion of the second frontal convolution. After the ventricle is entered the tumor mass usually can be seen bulging up into the anterio-medial portion of the ventricle. Again it is wise to insert a small blunt needle into the cyst rather than to make an incision directly through the presumed cyst wall with a

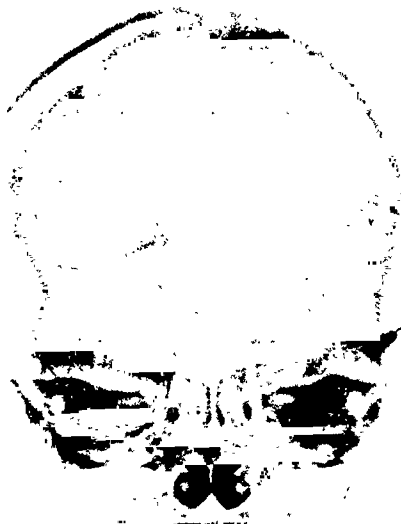


FIGURE 24. Roentgenograms showing air that had been placed into a craniopharyngioma cyst after aspiration of the cyst contents.

knife. After the cyst is entered, the technique for cleaning it out and for gradually collapsing the wall so that considerable amount of the tumor can be removed is identical to that described above. It is felt that all the neoplasm that is removable without precluding life should be removed.

Since all the craniopharyngioma cyst can seldom, if ever, be removed these tumors recur. The cyst fills up, often with material that can be aspirated through a No. 18 to No. 20 spinal needle. In these instances aspiration is accomplished after inserting the needle into the cyst through an open anterior fontanelle or a trephine hole. At the time of insertion of the needle roentgenograms may help in obtaining proper direction of the needle. The cyst can be reaspirated whenever it refills, usually every six to eight months. By this repeated tapping technique the size of the cyst, and concomitantly any increase in intracranial pressure can be controlled and these children remain relatively symptom free for periods of many years.

Occasionally sufficient tumor cannot be removed to decompress the third ventricle. The alternative then is to establish ventriculocisternal drainage (Torkildsen procedure). Obviously bilateral ventricular drainage is necessary.

Response of craniopharyngiomas to deep roentgen therapy has apparently been obtained in some clinics. There is no reason to believe the growth of the stratified squamous epithelial tissue is altered but deep roentgen therapy might attenuate the formation of cyst fluid. It has never been the policy in this clinic to suggest to the parents of the patient that deep therapy be tried.

If there is suppression of pituitary function by the tumor it is wise to start ACTH therapy four to five days prior to surgery and to continue it, but in decreasing dosages, for six to ten days postoperatively. An alternate method of protecting these children during the period of stress of the surgery is to administer cortisone. This should be started the day prior to surgery, giving 100 mgm. the day before surgery, 100 mgm. the day of surgery and also on the first postoperative day but then gradually tapering off the therapy over a period of six to eight days. Diabetes insipidus, that occasionally occurs postoperatively, usually needs no treatment for several days but if it continues longer, pitressin insufflations should be started.

Although tumors arising from the pituitary gland itself (chromophobe adenomas, etc.) are extremely unusual in children the technique of exposure is identical to that described above for craniopharyngiomas. Again it is emphasized that prior to incising the capsule of an adenoma, it is wise to insert a small needle to make certain there is no aneurysmal dilation of the carotid artery into the fossa. If the lesion proves to be an

adenoma the opening into the capsule is enlarged and the content curretted or sucked out. The prime endeavor is to remove sufficient tumor to decompress all the adjacent structures. These neoplasms are responsive to deep roentgen therapy—all should receive it postoperatively.

Pituitary Stalk Tumors

Tumors arising from the pituitary stalk *(infundibulomas)* are extremely rare. They were described first by Globus in 1942. He considered them to be of neurohypophyseal derivation and to originate from the saccus vasculosus in the infundibular stalk. Actually there is no saccus vasculosis in mammals but Globus postulated these tumors to arise from an abnormal arrest of this organ. The term infundibuloma was chosen because it best describes the derivation of the tumor as well as the characteristic vessel (the hypophyseal-portal veins) which are typical of both the tumor and infundibular regions of the human brain. Globus reported two cases, one in a 13½-year-old boy, the other in a five-year-old girl. Symptoms included headache, vomiting, dizziness, disturbed gait, and diplopia. The tumors extended into the floor of the third ventricle and produced hydrocephalus by involvement of the third ventricle with apparent blocking of the flow of fluid. The tumors are usually fairly cystic and can be rather vascular. The identifying histological characteristic is the numerous vascular channels typical of hypophyseal vessels. Around the vessels are numerous cells which have fairly large nucleoli with indistinct cytoplasm. Cellular elements are predominantly elongated unipolar or bipolar, not unlike the pituitocytes of the hypophysis or even bipolar spongioblasts. The nuclei of the fusiform cells are centrally situated. The protoplasmic processes taper off and are often undulated.

Pituitary Anterior Lobe Tumors

The anterior lobe of the hypophysis is formed from epithelial elements that evaginate upward from the buccal cavity. Tumors arising in this portion of the gland are fairly frequent especially in older people and have sufficiently specific clinical characteristics to be readily identified. However, in children, tumors arising in the anterior lobe are infrequent and comprise less than 1% of all the tumors seen in patients less than fifteen years of age. This infrequence is probably due to the fact that they are slowly growing and hence become manifest at a later age.

There are essentially four types of tumors arising in the anterior lobe: (1) *chromophobe adenomas;* (2) *eosinophilic adenomas;* (3) *basophilic adenomas,* and (4) *mixed type adenomas.*

Chromophobe Adenomas

These are the largest and most rapidly growing of all pituitary adenomas. The cells have no known secretory function and the tumors produce signs

and symptoms due to pressure on adjacent structures. On gross examination they appear soft, dark brown in color, and are usually well circumscribed. Occasionally they are invasive and extend up into the adjacent brain or they may erode into the sphenoid bone. On histological examination chromophobe adenomas may be extremely difficult or even impossible to differentiate from ependymomas. It has been reported that they actually are atypical ependymomas, a theory that cannot easily be cast aside. The chromophobe cells are polygonal and have fairly abundant cytoplasm. The nucleus is oval, dark staining, and at times pyknotic appearing. Mitoses are frequent. The vascular and connective tissue stroma is abundant and may divide the tumor cells into large groups or columns of cells.

As these tumors grow, they enlarge the sella producing a "ballooned out" appearance. Upward growth results in pressure on the diaphragma sellae resulting often in a "central or deep type" headache. After the tumor ruptures through the diaphragma the headaches may disappear or the headache may be replaced by one that is diffuse or occasionally by one located primarily in the frontal region. Further upward extension of the tumor results in pressure on the optic chiasm or optic nerves depending upon whether the chiasm is prefixed, postfixed, or normal in location. Visual disturbances especially defects in visual fields are the obvious clinical evidence of such pressure. The typical field of defect is a bitemporal hemianopsia. It begins as an upper temporal quadrant defect in each eye and proceeds downward in the temporal field. Occasionally a homonymous hemianopsia or scotoma may be present. Visual loss due to optic nerve atrophy may occur. Alpers in 1942 reported 79 patients with pituitary tumors. Fifteen (10%) were blind and 57 (71%) had a partial visual loss but his cases included patients of all ages. Pressure by the tumor on the secretory cells of the pituitary gland results in pituitary hypoactivity; in a child this is manifested by interference of growth (dwarfism) or occasionally by more severe hypopituitarism. In Alper's series, diminished visual acuity was the most frequent initial symptom, followed closely in frequency by headache. In fact, in one third of the patients headache was the initial symptom and occurred sometime during the illness in two-thirds of the cases.

Eosinophilic Adenomas

An eosinophilic adenoma produces hypersecretion of the anterior lobe. Occurrence of the tumor prior to the closure of the epiphysis results in "gigantism" and after the epiphyseal closure in acromegaly. These tumors are small, usually very slowly growing. They remain localized within the pituitary fossa; it is doubted whether they ever, in pure form, break through the diaphragma sellae to exert pressure on the visual system. Whenever there are visual disturbances associated with gigantism or acromegaly it

is considered due to a mixed type adenoma (chromophobe and eosinophilic). Histologically eosinophilic tumors are composed of polygonal or round cells with sharply defined boundaries. In the rather abundant cytoplasm are many fine acidophilic granules. The nuclei are round and vesicular. The cells lie in no particular arrangement in a sparse connective tissue and vascular stroma.

Basophilic Adenomas

Basophilic tumors are always small and restricted to the pituitary fossa. Often no definite tumor nodule can be found, but only an increasing number of basophilic staining cells, usually in the region of the pars intermedia. These tumors are presumably the cause of "Cushing's syndrome." *They are extremely rare; sufficiently rare even to doubt their occurrence* in children.

The treatment of chromophobe adenomas is primarily surgical for two reasons: (1) to verify the histological type of tumor, and (2) to decompress the optic chiasm and nerves. The concept that surgical attack on a presumed pituitary tumor is never justified until vision is threatened is considered wrong. *Since the pituitary tumors are extremely rare in children especially* when compared to other types of tumors occurring in this region (craniopharyngiomas, optic nerve gliomas, dermoids), which tumors may produce an amazingly similar symptomatology, absolute verification of histology can be accomplished only by biopsy. Sheldon has recently (1956) and wisely emphasized this concept. Surgical decompression should be followed by deep roentgen therapy. *Since these lesions are radiosensitive,* any recurrence can be treated with deep roentgen therapy.

The operative mortality in children is extremely low. Although the infrequency of these tumors in this age group have precluded valid statistics, it is surmised that the operative mortality would be 5% or less. The use of supportive hormonal therapy as outlined under the craniopharyngiomas *is recognized as an important adjunct in maintaining a low operative mor-*tality. Long term survivals can be anticipated, in fact, after surgical decompression plus deep roentgen therapy recurrence is not anticipated for at least three to five years. If the adenoma does recur, a repeat course of deep roentgen therapy is recommended.

Tumors classified as mixed type adenomas are treated the same as the *chromophobe adenomas The prognosis is essentially the same.*

A reasonably certain diagnosis can be made in most cases of eosinophilic adenomas. Since these lesions in pure form do not compress the chiasm or optic nerves the therapy, of course, is deep roentgen ray treatment. Only if there is no apparent benefit from the deep therapy is surgical interference considered justifiable The question has arisen whether the eosin-

ophilic adenomas are more sensitive to deep therapy than the basophilic; most observers believe the basophilic are more sensitive (Adelstein). It is universally accepted that chromophobe adenomas are least sensitive.

Pituitary Posterior Lobe Tumors

The posterior lobe of the hypophysis is composed of glial elements and is formed as a result of downward evagination of the floor of the third ventricle. According to Bucy (1945) only one authentic example of a tumor of the posterior lobe has ever been recorded and that was a ganglioglioma.

Optic Nerve Gliomas

At one time all tumors arising from the optic nerve were considered neuromas. According to Davis the earliest recorded example of an optic nerve "neuroma" was by Hegmann in 1842. In 1879, Willeman assembled from the literature twenty-five cases of optic nerve tumors of variable histology but most of them were gliomas. Verhoeff in 1921 reported on optic nerve tumors and stated that they were invariably gliomas. Davis wrote an excellent monograph in 1940 on primary tumors of the optic nerves. The majority were gliomas.

These gliomas are composed of astrocytes imbedded in a variable matrix of reticular and fibroglial tissue. They are comparable to the slowly growing pilocytic astrocytomas of the mesencephalon and pons and to the polar spongioblastomas in the region of the optic chiasm. They usually arise in the intra-orbital portion of the nerve although not infrequently the growth is restricted to the intracranial portion. There are no signs or symptoms that are entirely characteristic of optic nerve tumors. They must be differentiated from many other lesions occurring in this region. Usually symptoms begin during childhood or adolescence but can occur during later life. The history is often one of slowly progressive unilateral visual loss and painless exophthalmos with relatively slight limitation of ocular movement. On repeated ophthalmoscopic examination a progressively developing optic atrophy can be seen. Occasionally papilledema occurs but this is only after some primary visual loss. In the majority of cases there is roentgenological evidence of erosion of an anterior clinoid process or enlargement of an optic foramen.

When such a tumor is suspected, one should do a careful ophthalmoscopic examination, observe for alteration of extra-ocular movements, measure for any variation in pupillary size, and for exophthalmos, and test visual acuity. Consultation with an ophthalmologist is advisable. Skull roentgenograms should include special views of the optic foramina. It must be appreciated that not always are the foramina of equal size. Treatment is usually crani-

otomy with visualization of the region of the optic chiasm and nerves. The intra-orbital part of the nerve can be reached by perforating the roof of the orbit and enlarging the hole with a rongeur. Care must be taken not to perforate Tenon's capsule for then the fatty tissue of the orbit herniates through and tends to obscure vision. After the orbital roof is removed, the capsule should be incised in the direction of the nerve. The only structures to avoid are the ophthalamic branch of the trigeminal nerve and the branch of the oculomotor nerve that goes to the levator palpebral muscle. The amount of the tumor resected depends upon its appearance at the time of operation. Obviously if it extends back into the chiasm or over into the other optic nerve, simple biopsy followed by deep roentgen therapy is the procedure of choice. If the tumor is restricted to one optic nerve resection can be obtained and should be done irrespective of producing total visual loss in that eye. It soon would have been lost anyway. A decision on the extent of the surgical resection must be made at the time of exposure of the lesion. It must be remembered that deep roentgen therapy to these lesions is palliative and not curative.

Meningiomas

Meningiomas arising in the parasellar region are similar in appearance to meningiomas occurring elsewhere. Perhaps there is some greater tendency for them to be of a fibroblastic type than the more malignant types often found over the convexity. There was only one parasellar meningioma



FIGURE 25. Photograph of a 15-year-old girl with a left optic nerve glioma. There was total loss of vision as well as a 6 mm proptosis on the left.



FIGURE 26. Paraphyseal cyst in the third ventricle. These lesions are well circumscribed and surgically removable.

encountered in this series. In this patient the tumor had extended over against one optic nerve so there was diminished visual acuity, a minimal visual field defect, and unilateral primary optic atrophy. The tumor had also pushed upward into the hypothalamic region. There was mild diabetes insipidous, and, interestingly, pubertas praecox. Because of the location and extent of the lesion (with involvement of the carotid artery) total removal was impossible. Further discussion of the meningiomas is given elsewhere, the point here emphasized is that this tumor must be included in the differential diagnosis of lesions in the hypophyseal regions.

Mixed Tissue Tumors

These lesions are discussed elsewhere in this chapter. They occur primarily in the pineal region or cerebellopontine angle but occasionally they are encountered in the hypophyseal region. When they do occur here they lie just above the diaphragma sellae, immediately beneath the optic chiasm, and may extend upward around the chiasm. Preoperatively they cannot with certainty be differentiated from the craniopharyngiomas. Even the calcium present in the mixed tissue tumors is similar in radiographic studies to that observed in craniopharyngiomas. The surgical approach is identical to that of other lesions in this region. They are a bit less adherent than the craniopharyngiomas and total removal is possible. This, of course, is most desirable because these lesions are not sensitive to deep roentgen therapy.

Paraphyseal Cysts

Paraphyseal cysts are also known as *colloid cysts* of the third ventricle. They are very uncommon lesions and according to Shannon rarely appear before the age of twenty years. But the occasional one that does occur must be differentiated from a craniopharyngioma because of the different operative approach necessary to remove it and from the pineal tumors, because deep therapy does not affect the paraphyseal cyst. Interestingly these cysts can simulate symptomatologically either one of these two tumors.

The paraphysis was first described by Selenka in 1890 and is located in the anterior part of the roof of the third ventricle. Its function is unknown but some authors believe it is a vestigeal organ of hearing. In 1909, Sjovall thought remnants of the paraphysis might produce cysts and this is the generally accepted origin, although Zimmerman and German stated the cysts arise from the choroid plexus of the third ventricle because of their apparent attachment to the plexus as seen at the time of operation. On gross examination they are well circumscribed cystic tumors with a smooth translucent external surface. The capsule is composed of connective tissue the internal surface of which is lined with a layer of cuboidal or columnar cells. The cyst contains a homogenous gelatinous, liquid material.

These tumors lie in the third ventricle and cause symptoms due to cerebrospinal fluid obstruction as shown in Figure 26. They may produce intermittent headaches for a period of ten years although Dandy (1933) stated that 50% of all cases produce signs and symptoms of less than one year's duration. Perhaps the two most important clinical features of these lesions is the almost uniformly sudden onset of symptoms and the frequency of remissions. Zimmerman and German believe the ball-valve like action of the tumor in the third ventricle may account for these characteristics. A change in posture may shift the lesion giving relief of symptoms (similar to craniopharyngiomas). Stookey has stated that symptoms may arise from pressure by the tumor on the pyramidal tracts and nuclear masses adjacent to the third ventricle. This may cause periodic weakness of the legs and extraocular nerve palsies. Final diagnosis is made by ventriculography although these air studies may only show no filling of the third ventricle with dilation of the lateral ventricles. Occasionally one can visualize a dilated foramen of Monro with the rounded tumor projecting into it or, even less frequently, one can see a rounded, well circumscribed, ball-like tumor within the third ventricle.

If these tumors are recognized and treated early, a good prognosis is offered. Treatment is total surgical removal or if this seems impossible it is best to remove as much of the wall of the cyst as possible and permit the contents to drain into the ventricle. The cyst contents are not as

irritating as that from craniopharyngioma cysts. At the time of surgery these tumors are best approached by an incision into the ventricle through the mid-frontal convolution. The tumor can usually be seen bulging through the dilated foramen of Monro. A No. 22 needle should be inserted into the cyst and the capsule collapsed. When this is accomplished, it usually is possible to visualize two small arteries running from the choroid plexus to each side of the capsule. These should be clipped and cut following which the capsule can be delivered through the enlarged foramen. Occasionally it is necessary to enlarge the foramen. This is best done by an incision extending forward and upward from the foramen. The capsule usually is not adherent to the walls of the third ventricle.

Tumors in the Region of the Pineal Gland

Tumors in the region of the pineal gland are grouped together not only because they present characteristic symptoms and signs but also because they present a reasonably similar problem of therapy. Included in this group of tumors are the true pinealomas and the mixed tissue tumors that arise in the immediate pineal region, the gliomas (mostly astrocytomas) of the posterior third ventricular area, and the gliomas and cysts of the upper midbrain. A tumor that can mimic the above lesions is the angioblastoma that arises in the superior cerebellar region and extends up through the incisura into the midbrain. However angioblastomas are fundamentally cerebellar tumors and are discussed along with the posterior fossa lesions.

The clinical features most characteristic of tumors in the region of the pineal gland include abnormalities in function of the extraocular nerves (Parinaud's syndrome and/or strabismus) associated with evidence of increased intracranial pressure due to obstruction of the flow of cerebrospinal fluid. The various individual tumor types have, in addition to the above general symptomatology, identifying characteristics although it is oftentimes impossible to differentiate these various lesions clinically.

Pinealomas

Pinealomas are rare tumors occurring mainly in males under twenty-five years of age. They comprise about 2% of all gliomas. In 1943 Russel and Sachs reported on fifty-eight patients with pinealomas. They found that fifty-one (80%) of the fifty-eight were in males; twenty-eight (48%) were in patients between the ages of fifteen and twenty-five years and seventeen (29%) were in patients less than fifteen years of age.

The term pinealoma was first suggested by Krabbe in 1916. It is used to designate a tumor composed of two types of cells which in many instances show a type of arrangement suggestive of the mosaic pattern

observed in pineal tumors at the time of birth. The large cells possess a vesicular nucleus with a prominent nucleolus. The cytoplasm is relatively abundant. These cells represent the pineal parenchyma and have been described in detail by Berblinger. The small cells are lymphoid appearing and are enmeshed in a stroma composed of connective tissue cells with very elongated nuclei and of numerous stands of reticular fibers. An occasional plasma cell is present in the stroma. Some pineal tumors are composed of even more embryonic cells which resemble pineal proparenchyma. It was these tumors that Bailey (1932) called pinealblastomas, for at first he differentiated between pinealblastomas and pinealomas. Clinically it seems unnecessary to attempt this differentiation.

On gross examination pinealomas may vary from a smooth walled, fibrous, encapsulated mass with no invasive qualities to a highly invasive, necrotic, soft lesion that may spread along or into the ventricles and over the surface of the cerebrum and cerebellum or even invade the substance of the brain stem. Ectopic rests of pineal tissue apparently give rise to neoplastic growths and can occasionally be found in the midbrain and account for neoplasms in this region that have typical pinealoma or pinealblastoma structure.

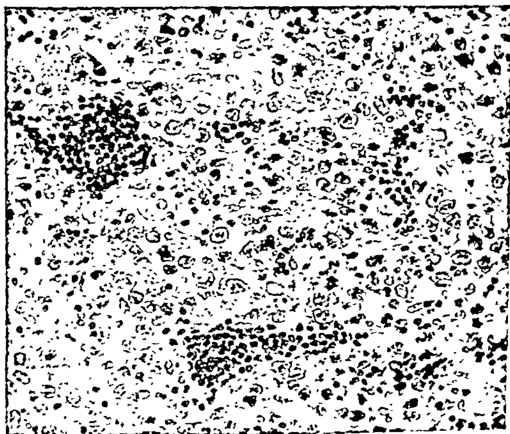


FIGURE 27. Photomicrograph of a pinealoma showing the two characteristic cell types—large cells with vesicular nuclei and small lymphoid appearing cells.

It has been recognized for many years that tumors occurring in the pineal region, and generally considered pinealomas, present a most varied clinical picture. Some of the tumors are well defined and circumscribed whereas others are diffusely infiltrative. Some respond well to deep roentgen therapy, others do not. Some are rapidly growing with a short clinical course, others are slowly growing with symptoms produced over a long period of time. To clarify this problem special studies have been made of tumors in this region. In 1944, Russel published a comprehensive monograph in which she concluded that many of the tumors heretofore considered pinealomas are actually atypical teratomas. It is now conceded that these two tumor types occur with about equal frequency in the pineal region. Often it is extremely difficult to differentiate them because of similarity of structure. But they should be differentiated to insure proper clinical care.

The question as to whether the pineal gland secretes a hormone that regulates sexual development has not been decided. Horrax and Bailey and others believe that the most acceptable theory of pineal function is that a secretion of this organ inhibits puberty. According to them an involutional change during puberty diminished its secretion thus allowing sexual characteristics and activities to occur. Others, including Russel and Sachs, do not believe it is a functioning endocrine gland. The syndrome of macrogenitosomia praecox is not common in these tumors but when present it occurs in males under the age of puberty.

There were ten patients with tumors diagnosed as pinealomas in this series. The average age was 11.4 years with a range from six to fourteen years. Eight were in males and two were in females. The average duration of symptoms were slightly over seven months with a range from two and one-half weeks to two years. All the tumors became manifest by headaches alone, by headaches and vomiting, or by headaches and deafness. Symptoms peculiar to pinealomas were due not to the type of tumor but rather to the location of the tumor. They arose in the pineal gland region and caused symptoms by direct invasion of the adjacent area or by pressure on the adjacent structures.

Perhaps the most suggestive localizing sign of a pinealoma is the inability to turn the eyes upward (Parinaud syndrome) and bilateral ptosis may be the first manifestation of this (Dandy 1933). Dandy stated that when ptosis is present bilaterally without further extraocular nerve palsies the findings are pathognomonic of a tumor pressing down onto the superior colliculi from the region of the pineal gland. Restricted upward gaze was present in eight of the ten children in this series. Bilateral hypoacusis and tinnitus may occur due to pressure on the inferior colliculi. The periventricular areas of the hypothalamus and basal nuclei were at times infil-

trated by the tumor and in one patient the tumor grew down into the pons and fourth ventricle. A symptom observed only in children with pinealomas was a peculiar spontaneous athetoid movement of the arms and hands. Other symptoms that occurred predominantly in these tumors were polyuria and polydypsia, rapid body development, tinnitus and deafness, and mental dullness. None of the patients showed evidence of precocious puberty but all had definite metabolic disorders. They appeared ill and had a multiplicity of complaints. Very frequently there was involvement of the long motor tracts manifested by weakness in the extremities and reflex changes. This was the only type of tumor in which patients showed a gain in weight although paradoxically many appeared more emaciated and anorexic than the average patient with a brain tumor. There may also be signs of apparent cerebellar involvement such as staggering gait, etc. Globus and Silbert reported cerebellar signs in four of seven cases. Signs of increased intracranial pressure due to obstruction of the aqueduct occur often and Bailey *et al.* (1939) stated that precocious puberty and general overdevelopment associated with increased intracranial pressure should always make one suspect a pineal tumor. Unusual calcification of the pineal gland is suggestive of a pineal tumor but this is not a trustworthy sign. In only two patients was there calcium, visualized in roentgenograms, of sufficient amounts to be considered diagnostic of a pineal neoplasm. Pineal tumors may simulate posterior fossa lesions. This was true in this series because of the early signs of intracranial hypertension, stiff neck, positive Romberg test, trunkal ataxia, and even past pointing. Ventriculography may be necessary to demonstrate these lesions adequately and Bailey *et al.* advised this in all cases in which the characteristic eye signs were absent.

The problem of surgical management of tumors in the pineal region is quite dependent upon the histology of the tumor. Obviously the pinealomas are seldom if ever small discrete lesions that can be removed. More likely their infiltrative character, in addition to their location, makes surgical removal impossible. If direct surgical removal of the tumor is to be attempted, the most accepted operative approach was described by Brunner and modified by Dandy in 1921. It consists of going down along the falx, splitting the splenium of the corpus callosum, and entering the pineal region. It was not until 1931 that Dandy had his first apparent total removal of a pineal tumor but this patient later had a recurrence of the tumor. In 1936, he reported removal of tumors in this region in three patients. One was a teratoma; this patient died in four months. Another died in two and one-half years from a recurrence and the third was living 4 months after removal of the pinealoma. He reported as complications of this approach, in addition to inability to remove the tumor, extraocular muscle palsies and transient blindness due to an edema of the quadri-

geminal plate. Kahn reported upon a pinealoma totally removed by Peet in 1929 and a teratoma by himself in 1935. He stated that with the exception of these two cases no patients in which extirpation was attempted by him survived sufficiently long to warrant the operative procedure. Others (Horrax and Bailey, Dandy) have reported similar experience. Of the ten cases included in this report, in only two was direct operative attack made and in both it was unsuccessful. It is now the policy to carry out ventriculographic and possibly angiographic studies on these patients suspected of harboring a tumor in the pineal (posterior third ventricle) region. Only in those patients in whom there is seen a discrete, well rounded lesion protruding downward and anteriorly into the third ventricle and in whom the suprapineal recess is visualized is a direct attack on the tumor considered. Lesions with this radiographic appearance have a good chance of being a teratoma, simple cyst, or aneurysm, and hence not responsive to deep roentgen therapy. However, even in them direct operative attack has not been particularly spectacular as far as results are concerned. Very few patients have ever been improved sufficiently to warrant this approach. It is believed a better method of therapy in patients with teratomas in the pineal region is to perform either a subtemporal decompression or one of the cerebrospinal fluid shunting procedures. It is considered immaterial whether the shunting procedure is that described by Torkildsen or a puncture of the lamina terminalis. The latter has the advantage of not needing open subarachnoid spaces around the pons to afford a channel for the upward flow of the fluid. Consequently, when such a block is suspected by the air studies a third ventriculostomy (puncture of lamina terminalis) is carried out. However after doing a lamina terminalis puncture it is not unusual for the subarachnoid spaces in the cisterna chiasmaticus to become closed by reaction to blood from the operative procedure so that a functioning shunt is not obtained. Because of this most surgeons prefer the Torkildsen procedure. If the cisterna around the pons is obliterated by the tumor, the Torkildsen procedure can be combined with an incision into the tentorium to enlarge the incisura. Obviously, however, this is a technically difficult procedure and adds considerably to the mortality and morbidity.

If the tumor is considered a true pinealoma because of its clinical characteristics and its appearance in ventriculography a shunting procedure is carried out and the patient then given deep roentgen therapy. Occasionally a subtemporal decompression is performed rather than the shunting procedure. However, the subtemporal decompression is generally considered inferior to the shunt for control of pressure due to obstruction of the aqueduct.

The recommended dosage of radiation is 4000 to 5000 roentgens tissue

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dosage given through multiple ports over a period of four weeks. Because the child is so uncomfortable and often sufficiently ill to necessitate intravenous feeding and other supportive therapy it always is wise to keep the child hospitalized during the first two weeks of radiation therapy. Thereafter it can usually be given on an "out-patient" status.

Prognosis in children with pinealomas is not too bad. Following deep roentgen therapy symptoms generally resolve and a fairly normal life can be led. Recurrence may, of course, come at any time but usually a period of two to five years supervenes at which time the deep roentgen therapy can be repeated. Occasionally the period of regression or freedom from symptoms is much greater than five years, occasionally ten to fifteen years.

Mixed Tissue Tumors in the Pineal Region

The teratoma is the usual type of mixed tissue tumor occurring in this region. A detailed account of them has been published by Russell in 1944. She believed that teratomas, or some variation thereof which she considered atypical teratomas, compromise about 50% of all tumors in the pineal region. This seems unusually high, especially when the tumors are limited to those in children. In this series of 273 tumors there were eleven pinealomas but no teratomas. Weber reported on teratomas occurring

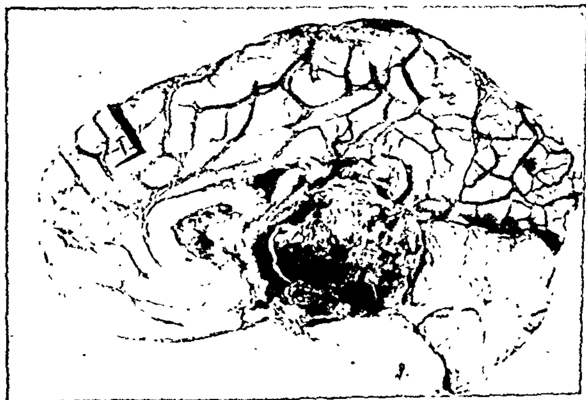


FIGURE 28. Teratoma, diffuse and infiltrative, in the region of the pineal gland. Surgical excision obviously was not possible.

irrespective of age and found 53% of them to be in the pineal region, but in this group reported by Weber there were very few children.

The general characteristics of mixed tissue tumors are given elsewhere in this chapter (page 294). The only point of emphasis is that whenever a pineal region tumor is suspected, a ventriculogram should be done and if the tumor appears discrete, well rounded and well defined, a diagnosis of a teratoma, rather than pinealoma, must be strongly entertained. The problem then arises as to the best form of treatment. These are slowly growing, developmental type tumors. It is believed a shunting (Torkildsen) procedure should be done. If this controls the symptoms no further therapy is feasible but if disabling symptoms persist an attempt at direct surgical removal or at least intracapsular decompression is advised. The possibility of obtaining total removal is remote as suggested above by Kahn. Therefore the intracapsular decompression is usually all that can be done.

Simple Cysts

Simple cysts are unusual lesions that occur predominantly in the upper pons. They can simulate clinically pineal tumors and hence are included here. In 1942, Globus, Kuhlenbeck, and Weller reported on tumors of the aqueduct of Sylvius, and found two lesions that fell into the category of simple cysts. These lesions must be differentiated from the well known porencephalic cyst. The simple cysts, of which there was one in this series of 273 cases, probably arise as a result of congenital malformation although one can presume they are really angioblastic lesions in which the minute mural nodule is not found. These cysts appear as small cavities filled with clear fluid and have a smooth lining membrane of fibrous tissue or occasionally a layer of cuboidal cells. They should be differentiated from the malformations of the aqueduct described by Russel.

Symptoms produced by simple cysts, which occur mostly in the pons, are due to block off the flow of cerebrospinal fluid. Obviously they are difficult to diagnose and the majority of those reported in the literature were found at autopsy in patients suspected of having a brain stem glioma. If a simple cyst lesion is suspected, surgical removal obviously is impossible. A ventriculocisternal shunt (Torkildsen) is considered the most appropriate treatment. If the cyst is of sufficient size to enlarge the pons to the extent the subarachnoid channels around the pons are obliterated, either the tentorial notch must be enlarged or a third ventriculostomy (Scarff) can be done.

Gliomas in the Pineal Region

Any of the various histological types of gliomas may occur in the pineal region. In this series of 273 tumors in children there were five astrocytomas,

three astroblastomas, three spongioblastomas, and one glioblastoma. Ordinarily one would expect in a series of this size to encounter a few ependymomas but this, for some reason, did not occur. The gliomas in the pineal region produces symptoms very comparable to those of a pinealoma. The only difference is that growth disturbances and perhaps Parinaud's syndrome was less frequent whereas abnormal dystonic (athetoid) movements were more frequent. Additionally, the involvement in the gliomas was more apt to be unilateral than that observed in the more midline located pinealomas. But differentiation often cannot be made on clinical grounds. During the routine investigation of a lesion in this area air studies are quite invariably performed and a clue to the fact that the lesion is a glioma may be obtained. But then what is the proper therapeutic approach? It is usually unwise to attempt surgical removal because these lesions are diffusely infiltrative and very little can be gained by such an attempt. Even to biopsy the lesion is hazardous, but this may be necessary to verify the diagnosis. After this has been accomplished, and if an obstruction to the flow of cerebrospinal fluid is present a shunting procedure (Torkildsen) should be done. Deep roentgen therapy should then be administered to the lesion. The approved dosage is at least 4000 tissue roentgens given through multiple portals, in divided dosages over a period of four to five weeks. Occasionally these gliomas are very responsive to deep roentgen therapy, in one patient symptoms remained in abeyance for nine years.

Miscellaneous Tumors

Tumors considered under this group include the meningiomas, the tumors of mixed tissue, metastatic tumors, chordomas and chordoblastomas, and granulomas. They are placed together for discussion primarily because they do not fit into any other of the groupings—this is their only common factor. Some of them, such as the meningiomas and the metastatic tumors may be multiple, none have a single or invariably consistent site of origin except the chordomas.

Meningiomas

Meningiomas comprise about 5% of intracranial neoplasms in children, a much lesser frequency than observed in adults. Cushing in 1927 found only two in a series of sixty-two children with brain tumors but Bailey *et al.* reported eight in one hundred verified brain tumors. There were only seven children with meningiomas in this series of 273 verified brain tumors.

Meningiomas arise from the cap (meningiothelial) cells of the arachnoidal granulations. The arachnoidal cap cells are normally well differentiated cells. In meningiomas this differentiation persists and these arach-

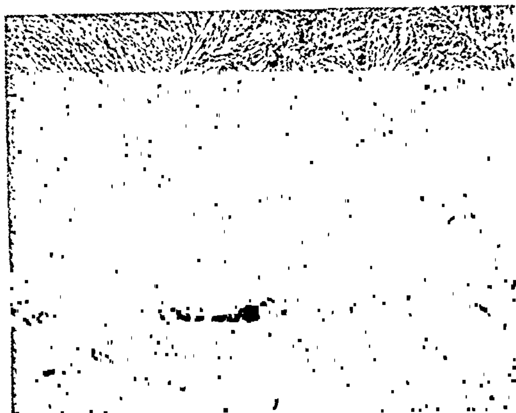


FIGURE 29. Photomicrograph of a fibroblastic meningioma. This type of meningioma is, unfortunately, seldom seen in children. They are firm, usually discrete and surgically removable tumors.

noidal cap cells are found enmeshed in a supporting and nutritive stroma composed of several elements, the most constant of which are collagenous connective tissue and blood vessels. At times there is an overgrowth of the supporting stroma and the original cells from the arachnoid are overlooked. A fibroblastic type meningioma, for example, is composed almost entirely of the proliferative supporting stroma, the true meningotheliomatous cell may not be discerned.

Usually meningiomas are localized, well circumscribed lesions, but certain types exhibit definite malignant characteristics. The latter is especially true in children. Whereas in adults meningiomas represent one of the more prognostically favorable types of brain tumors, in children they are often inoperable. They tend to be histologically malignant; there is a preponderence of the more rapidly growing sarcomatous, angioblastic, and mesenchymal types, and very few of the fibrous, psammomatous, and meningotheliomatous types. Ingraham and Scott have stressed the fact that meningiomas in children tend to be malignant, and Bailey *et al.* also stated that they are apt to be "more embryonic, more rapidly growing, with invasive tendencies so that they might better be called sarcomatosis." Often there are multiple sites of origin. These more malignant meningiomas in children should be differentiated from the malignant dural tumors of

purely connective tissue origin, that is, fibrosarcomas of the dura. The fibrosarcomas of the dura have been divided by Bailey and Ingraham into a diffuse saccular type and a localized solid type. The former are extremely malignant, often bilateral, and are considered hopeless in prognosis. The latter are more localized, well defined and at times are surgically removable.

In the series of seven meningiomas in children observed at this clinic, two were lipomatous, two psammomatous, one angioblastic, one sarcomatous, and one meningothelial in type. The meningiomas were supratentorial in five patients and subtentorial in two. In three patients the tumors were multiple. The general prognosis was poor as demonstrated by the fact that all cases terminated fatally within a month after operation except one who has lived four and one-half years.

There are no characteristic features of the symptomatology or of the signs that might make one suspect preoperatively the child has a meningioma unless the lesion is suspected as being one of the various manifestations of multiple neurofibromatosis (von Recklinghausen). This was true of two of the patients in this series, but the others were encountered without foresight that a meningioma was present. In one of these patients, it was suspected preoperatively that a subdural hematoma was present. This was a six-month-old child with vomiting, enlarged and bulging anterior

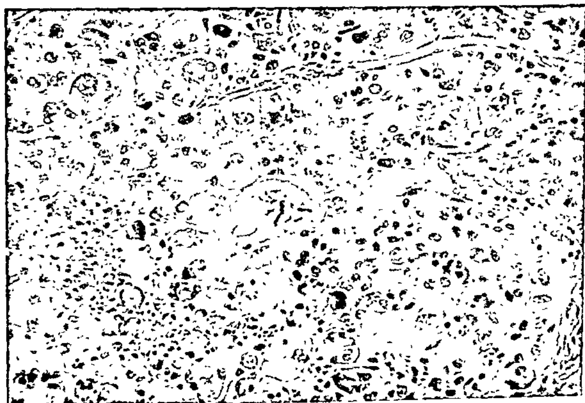


FIGURE 30 Photomicrograph of a malignant sarcomatous type meningioma fairly characteristic of meningiomas in children.

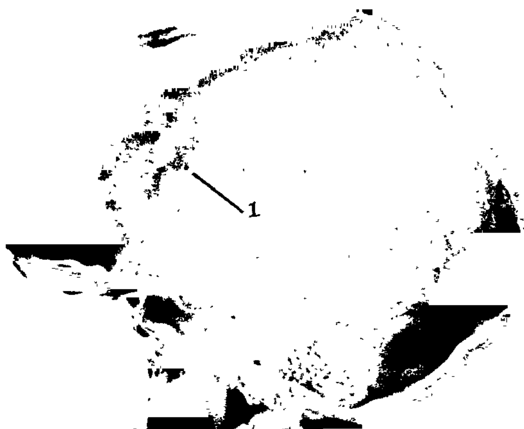


FIGURE 31. *(Top)* Roentgenogram showing calcium in a lipoma arising from the anterior part of the corpus callosum. These are invariably inoperable tumors.

FIGURE 32 *(Bottom)* Roentgenogram showing calcium in large cholesteatoma. The bulk of the tumor was at number 2 but there was a cyst with calcified wall at number 1.

fontanelle, and increased circumference of the head. At the time of surgery the "hematoma" was removed. This "hematoma" consisted of a rather thick and shaggy inner and outer membrane within which was chocolate colored fluid. The "hematoma" membrane was reported histologically to be a sarcomatous meningioma. In retrospect it more probably was a fibrosarcoma of the diffuse saccular type. Obviously no cure was accomplished. The case was seen prior to the time that angiography became a frequent diagnostic procedure. Even so it is believed that at present the diagnostic subdural taps via the anterior fontanelle would have been done in this age child—and an erroneous preoperative diagnosis again made. In retrospect a hint to the correct diagnosis was that the fluid aspirated was more brown and semigelatinous in character than that obtained from the usual subdural hematoma.

The variable characteristics of meningiomas in children makes an adequate discussion of their surgical management impossible. When they are located in the posterior fossa the most probable location is in the cerebellopontine angle and the surgical approach is comparable to that described for acoustic neuromas. When they are located in the supratentorial region there is no characteristic site of occurrence nor is there any particularly auspicious or encouraging method of management. Since meningiomas may be multiple, invasive, and large it is well to be fortified preoperatively with all the surgical adjuncts available. These include adequate amounts of cross matched blood and "hypotensive anesthesia." Unfortunately "hypotensive anesthesia" is not as adaptable or proficient in children as in adults. Extremely important is a thorough knowledge of the location and approximate size of the arterial supply to the tumor. If it is at all possible it is wise to occlude these vessels just proximal to the tumor prior to any attempt to enucleate the lesion. In order to remove the lesion totally, it is necessary to excise the involved dura. This leaves a defect or hole in the dura. This defect in the dura should be covered by a flap constructed by separating off the outer layers of the adjacent dura and turning this flap back as a hinge over the defect. If this is not possible because of the size of the defect, temporal fascia, periosteum, or polyethylene film can be used. Involvement of the overlying bone by a meningioma in a child has not been observed in this clinic, if it is involved it should be removed and a cranioplasty done.

Tumors of Mixed Tissue: Epidermoid, dermoid, and teratoma

Tumors of mixed tissue can be classified into three types depending upon the number of germ layers present. The *epidermoids* are composed of elements of one germ layer, the ectoderm; the *dermoids* are composed of elements of two germ layers, the ectoderm and the mesoderm; the

teratoma is composed of elements representing all three germ layers. There is still controversy concerning the origin of these tumors that obviously arise from embryonic rests. Hosoi has suggested that a tumor that is obviously monodermal may have been tridermal in the beginning but one cell type grew rapidly thereby overshadowing other types. Another postulate is that the age of embryonic life at which the inclusion occurs determines the structure of the resulting tumor. An inclusion in early embryonic life causes the isolation of multipotential cells that are capable of forming hair and glandular structures. Inclusions at a later date involve cells which already have differentiated and so development is limited to simple squamous epithelium. Love and Kernohan have ably discussed these lesions in 1936.

EPIDERMOIDS (CHOLESTEATOMAS)

There were four patients with epidermoids in this series; in one it was located in the cerebellopontine angle and in three they were in the suprasella region. These two sites seem to be the most typical locations. Another frequent location is within the fourth ventricle or right at the foramen of Magendie. When epidermoids do occur there is nothing in the clinical features to differentiate them from other slowly growing brain tumors. The average duration of preoperative symptoms in this series was seven years with a range from five to eleven years. In all cases the initial symptoms were headache, vomiting and loss of vision. Three of the patients were blind when first seen in this clinic—all three had suprasella epidermoids. In two patients there were associated developmental anomalies—an asymmetrical face with a cleft lip in one patient and a dermoid cyst on the tip of the nose in the other. Because these tumors probably arise as inclusions, an epidermoid must be considered whenever a midline lesion is suspected. These tumors calcify and this may be a clue to their presence. Mahoney stressed the great frequency of mental symptoms in patients with mixed tissue tumors and this has been observed and discussed in this clinic too.

The method of treatment is surgical excision of the tumor mass. Seldom are they adherent to the surrounding brain; total removal is usually accomplished. At times one must be satisfied with an intracapsular removal but the tumor will recur if the capsule is left behind. It may take several years before the recurrent tumor becomes manifest. At the time of excision care must be taken to avoid spilling the contents of the tumor into the ventricle or the subarachnoid space. A sterile type meningitis may result from the chemical action of the cholestrin. Also debris from the sacs of these tumors may get into the ventricular system and plug the aqueduct.

DERMOIDS

Dermoid tumors are less frequent than epidermoids. There was only one in this series—it was located in the temporal lobe. The favorite site of location is along the midline, usually at the base of the brain or in the region of the fourth ventricle. There are no signs nor symptoms entirely characteristic of dermoid tumors. They are slowly growing lesions and may give symptoms over a prolonged period (5-10 years). Occasionally these tumors are multiple; they may be associated with other congenital anomalies; at times the capsule is calcified and can be seen in roentgenograms.

Treatment is surgical removal; deep roentgen therapy is of no avail. It is emphasized that at the time of surgical excision care should be taken not to spill any of the contents of the tumor for fear of a chemical meningitis with later local scar formation. The one patient in this series with a dermoid tumor had total removal of the lesion but in this instance the tumor was small, well circumscribed, and easily removed. Obviously not always can the surgeon, and patient, be so fortunate.

TERATOMAS

The sites of predilection for teratomas are the pineal and pituitary regions but they occur in many other places too. Weber found in a series of sixty intracranial teratomas that thirty-two (53%) were in the region of the pineal gland, twelve (20%) in the region of the hypophysis, four (6%) in the lateral ventricle, and two (3%) were in the orbit. The remaining ten cases were located in various sites throughout the cranial axis. In the University of Minnesota series there were only two patients with teratomas —in both the tumor was located in the suprasellar region. There seems to be some sex predilection with teratomas occurring twice as frequently in boys as in girls.

There are no symptoms characteristic of teratomas except for the typical sites of occurrence in the pineal or in the suprasellar region and the fact that they are very slowly growing lesions that produce symptoms of several years duration prior to the patient presenting for surgery. Teratomas are not particularly invasive tumors; symptoms caused are those due to pressure on adjacent areas. Dandy stated that when these tumors occur in the pineal region the most significant symptom is restricted upward gaze (Parinaud syndrome) and that the earliest objective evidence of this symptom is a bilateral ptosis. He believed that whenever ptosis was present bilaterally without further extraocular nerve palsies, the findings are pathognomonic of a tumor pressing down upon the corpora quadrigemina from the region of the pineal gland.

The treatment of teratomas is total surgical removal since, if incompletely removed, the tumor recurs. This often can be accomplished in

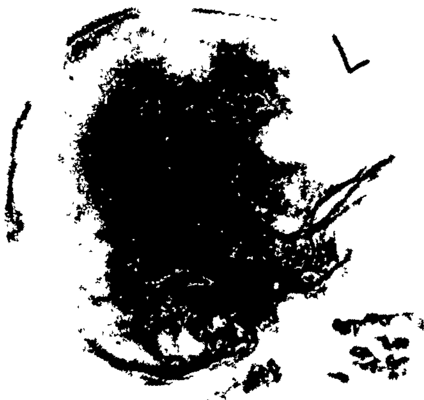


FIGURE 33. *(Top)* Photomicrograph of teratoma showing structures arising from all three germ layers.

FIGURE 34. *(Bottom)* Roentgenogram of child, age eight, with a neoplasm metastatic to the calvarium This was a Ewing's tumor. It had involved numerous bony structures throughout the body.

those located in the suprasellar region but the outlook is not so fortunate in those in the pineal region. Seldom can these be removed totally and all that can be hoped for is an intracapsular decompression. Recurrence is inevitable and becomes manifest usually in one to three years. Deep roentgen therapy is of no avail. If it is deemed inadvisable to attempt direct attack on those tumors located along the midline in which there is a block of flow of cerebrospinal fluid, a Torkildsen procedure to shunt the flow of fluid around the obstruction is advisable.

Metastatic Brain Tumors

Tumors may metastasize to the brain or calvarium from anywhere in the body. In children the most frequent site of the primary is the adrenal gland; the next most frequent is the kidney. Melanomas in children also not infrequently metastasize to the brain. In this series of 273 patients there were twenty-two (8.0%) patients with metastatic brain tumors. In four the metastases seemed to be solitary lesions and were located in the cerebellum and in four the apparently solitary lesions were in the frontal lobe. All the others were multiple. There were no characteristic symptoms nor signs indicative of these lesions being metastatic except in those cases in which there was a known primary. In two cases an increased blood sedimentation rate without any evidence of infection was a clue to the lesion being metastatic. In both of these cases there were multiple metastases. A routine preoperative diagnostic procedure should be a chest roentgenogram. The same is true for a blood sedimentation rate.

The problem exists of whether or not surgical excision should be done in a patient with a known metastatic lesion. The policy followed in this clinic is to excise the intracranial metastasis if it seems the metastatic lesion will cause death or severe neurological dysfunction even though the patient's general condition is otherwise reasonably good. An exception to this is the child in whom the intracranial pressure is not rapidly progressive and the tumor type is radiosensitive. Deep roentgen therapy should be administered. Multiple intracranial metastases are best treated with deep roentgen therapy—surgery on one of the metastases may provoke a progressive and fatal edematous reaction around the other lesions. In order to tide the patient over during the course of deep roentgen irradiation, a subtemporal decompression may be necessary. Perhaps the best way to determine whether or not a subtemporal decompression is necessary is to follow the patient's general systemic condition and to obtain visual acuities every other day. An appreciable deterioration in the patient's general condition or a decrease in visual acuity is believed to be an indication for immediate decompression The type of decompression done is the myoplastic procedure described by *Penfield* and *Cone* plus the *dural splitting* technique of Elsberg



FIGURE 35. Photomicrograph of a chordoma showing masses of large epithelial cells.

Chordomas and Chordoblastomas

Chordomas and chordoblastomas are extremely rare tumors that become symptomatic most frequently in later life. There are only a few reports of these tumors occurring in children. They are mentioned here primarily for consideration in differential diagnosis of lesions involving the brain stem. These tumors have their origin in the basisphenoid region from the remnant of the notochord known as the ecchondrosis physalifora. On gross examination the tumors have a nodular but smooth capsule of a milk-white color. The cut surface is translucent and mucinous in consistency. Brown streaks due to hemorrhage may be present, although these tumors are usually poorly vascularized. On histological examination they are composed of cords and masses of large epithelial like cells. The cells are sharply delineated and have a dark eccentrically placed nucleus surrounded by large amounts of vacuolated cytoplasm. They are very slowly growing lesions that may remain small and asymptomatic or become large and produce symptoms by involvement of the cranial nerves in the forward part of the posterior fossa. The pons may even be distorted by the tumor. Roentgenograms may reveal a local destruction of the sphenoid bone—a finding characteristic of these tumors.

a rapidly growing neoplasm but that it is only a slowly growing neoplasm that will produce symptoms for a long duration. Likewise it is emphasized that there is a great variability in the duration of symptoms produced in an individual by any one of the various tumor types and these data can be used only as suggestive evidence for differentiation of these lesions.

Location	*Number of Cases*	*Preoperative Symptoms*
Cerebellum		
Midline	61	16 8 months
Hemisphere	37	16 6 months
Brain Stem	24	8.1 months
Cerebellopontine Angle	6	7 2 years

FIGURE 37. Comparative duration of symptoms in patients with tumors in the posterior fossa.

Tumors of the Cerebellum

The majority of tumors arising in the cerebellum in children are gliomas of which the astrocytomas, the medulloblastomas, and the ependymomas are the principle histological types. Also occurring, but relatively infrequently, are sarcomas, angioblastomas, metastatic tumors and granulomas. For purposes of therapy it is important to differentiate these histological types. Unfortunately, with the exception of the metastatic tumors and granulomas, there is no particular way of differentiating them preoperatively and surgical biopsy is necessary to establish the histological type. The concept that accurate differentiation is possible by purely clinical means should be condemned. There are no reliable preoperative criteria for differentiation and surgical biopsy definitely is necessary to establish the histological type. It is inexcusable to give deep roentgen therapy to a child with a midline cerebellar neoplasm without histological verification; recognition that the lesion is not a medulloblastoma but a surgically curable astrocytoma may come too late—after the child is blind.

Midline Cerebellar Tumors

Midline cerebellar tumors arise either in the vermis or from the walls of the fourth ventricle. Symptoms produced by them are due either to obstruction of the flow of cerebrospinal fluid or to dysfunction of the vermis. There may be secondary involvement of the cerebellar hemispheres or brain stem. The most frequent symptom is vomiting and this occurs in about 90% of all these patients Headache is the next most frequent symptom and occurs in about 75% of all these patients. Headache is usually localized over the occiput, and it is frequently accompanied by pains in

the posterior cervical region or by stiff neck. However, the headache may be localized over the eyes or forehead; this should not lead one away from consideration of a cerebellar neoplasm. There is unsteadiness in walking or sitting (trunkal type ataxia) in about 60% of the patients with midline cerebellar neoplasms whereas incoordination of the arms is quite unusual. It is emphasized that a definite incoordination of the arms is very suggestive that the lesion is located in the cerebellar hemispheres and not primarily a vermis lesion. Further symptoms include listlessness and somnolence, personality disturbance, convulsive seizures (cerebellar seizures, generalized seizures, or even focal seizures), failing vision from papilledema or second optic atrophy, and diplopia.

As a midline cerebellar tumor enlarges it soon fills up the fourth ventricle. The flow of cerebrospinal fluid is blocked and signs characteristic of increased intracranial pressure become evident. In younger children there is enlargement in the circumference of the head with bulging fontanelle. Oftentimes papilledema is present even though the enlargement of the head is an attempt to compensate for the increased pressure. In older children papilledema is almost always present. In addition to the evidence of increased intracranial pressure, signs of involvement of the vermis become apparent. Principal among these are a trunkal type ataxia with a positive Romberg test but with minimal incoordination on doing the finger-to-finger, finger-to-nose, and finger searching tests. The nystagmus, which is present in about 50% of all patients with midline cerebellar tumors, is usually a bilateral horizontal type nystagmus although occasionally a vertical type may also be present. Characteristically the deep tendon reflexes are hypoactive and there is some hypotonicity of the muscles. Usually in children with neoplasms of the midline cerebellar region these changes are present bilaterally but in about 10% of children unilateral reflex abnormalities are found even though the lesion is midline. Occasionally the deep tendon reflexes are hyperactive. This may occur in a patient with either a midline or laterally placed cerebellar neoplasm and is considered a result of pressure onto or direct tumor infiltration into the long motor tracts in the brain stem. The great toe signs (Babinski) are positive bilaterally in about 15% and unilaterally in about 10% of patients with cerebellar tumors. Suboccipital tenderness is frequent in patients with cerebellar tumors either in the midline or hemisphere. When it is obtainable only on one side it is very suggestive that the tumor underlies the tenderness. Occasionally a patient with a midline cerebellar tumor holds his head rigidly, always trying to prevent any flexion or rotation from taking place because pain from pressure by the tumor onto the upper cervical or lower cranial nerves follows any movement of the neck.

Tumors in the Cerebellar Hemisphere

Symptoms produced by neoplasms in the cerebellar hemispheres are due: (1) to the obstruction of the flow of cerebrospinal fluid with a resulting increased intracranial pressure; or (2) a dysfunction of the cerebellar hemispheres or the afferent or efferent pathways to the cerebellar nuclei. Vomiting is the most frequent symptom and occurs in about 90% of these patients. Headache is the next most frequent symptom. Also occurring are listlessness, somnolence, change in personality, and ataxia involving primarily the extremities. This incoordination of the arms and legs is one of the most valuable adjuncts in differentiating a cerebellar hemisphere from a midline cerebellar neoplasm because the ataxia observed in a patient with a midline cerebellar neoplasm is trunkal in type and usually there is very little if any involvement of the arms or legs. Similarly tremor of an extremity occurs frequently in patients with cerebellar hemispheral tumors and only rarely in those with midline lesions. The incoordination and tremor, of course, are ipsilateral to the involved hemisphere. As a general rule hemispheral cerebellar tumors are larger than midline lesions and are more apt to cause pressure on the brain stem structures with long motor (pyramidal) tract or cranial nerve involvement.

The signs produced by a tumor in a cerebellar hemisphere are usually very characteristic. These include evidence of increased intracranial pressure, i.e., bilateral papilledema, occasionally oculomotor or trochlear nerve palsies, enlarged circumference of the head, widening of the cranial sutures, and increased cranial digital markings. Of course, not all these signs may be present in any one child. *Signs specifically indicative of hemispheral involvement include ataxia involving one arm or leg and, of course, this* ataxia is on the side of the lesion. The Romberg test is usually positive and there is a tendency for the patient to fall toward the lesion. If the patient attempts to walk with closed eyes, he veers toward the involved hemisphere. Past pointing occurs on the side of the lesion and a combination of unilateral tremor, past pointing, incoordination of the arms with a positive Romberg sign and a tendency to fall or walk to one side is almost pathognomonic of a laterally placed cerebellar tumor. Conspicuous by their absence or great infrequency in patients with tumors of the cerebellar hemispheres are visual field defects, primary optic atrophy, cranial nerve palsies other than the abducens, oculomotor and trochlear nerves, paresis or paralysis of an arm or leg, and abnormal body growth.

Surgical Consideration of Cerebellar Tumors

The sitting-up position is used in all posterior fossa explorations except in very young patients (one year old or less) with fairly extensive hydro-

cephalus. In these instances, with the patient in the sitting position, fluid escapes through the posterior fossa exploration and a massive collapse of the very thin cerebral mantel may occur. The fragile veins coursing between the cortex and the meninges may rupture resulting in the formation of a subdural hematoma. Concomitantly air that has gotten up over the cerebral hemispheres has been known to enter these torn veins. The danger of an air embolus is ever existent! Both these complications have been experienced so that a strict policy has been instituted that no infant with appreciable hydrocephalus is operated upon while in the sitting position. When the sitting position is not used the patient is placed face down using a horse-shoe shaped head rest with a small roll or a brace elevating both shoulders and hips permitting easy excursion of the abdomen and chest with each compression of the anesthesiologist's rebreathing bag. The important feature is that the patient must be placed in a position so that the cardiorespiratory status can be readily surveyed and controlled by the anesthesiologist.

It is important to prepare and drape the patient in such a manner that a trephine hole can be placed in the right and left occipital areas thereby permitting one to decompress the lateral ventricles. Likewise it is necessary to drape out laterally to the mastoid region and then down over the cervical vertebrae, extending down as far as the fifth cervical vertebrae (Figure 38). The latter is necessary because tumors not infrequently extend down this far, and it is very dangerous to change the position of the drapes after the skin incision is made and exposure has been obtained.

Routinely a midline skin incision is employed. The occipital muscles are separated subperiosteally from the occipital bone and the erector capiti muscles from the first, second, and third cervical vertebrae. The occipital muscles are detached along the superior nuchal line bilaterally for a distance of about three centimeters. By so doing very adequate exposure of both the right and left cerebellopontine angles can be obtained, especially in children under six years of age. In older children the midline incision is also used and only under extremely unusual circumstances is the incision carried in a hockey stick manner out laterally to the mastoid. If one is using the sitting-up position it is imperative that a gauze sponge be kept in the line of the incision to prevent any passage of air into the veins prior to coagulation of these vessels. After the occipital bone is exposed adequately a burr hole is made into the bone on both sides of the midline. Then bone is rongeured away superiorly to the lateral sinus, laterally to the mastoid process, and downwards into the foramen magnum. If the neoplasm is located in one cerebellar hemisphere the bone is not taken off on the contralateral side unless it is absolutely necessary in order to obtain adequate exposure. The opening into the foramen magnum is

carried out laterally as far as possible without risking an injury to the vertebral vessels. The arch of the first cervical vertebrae always is removed and if necessary the laminectomy is carried down to include the second and third cervical vertebrae. For unilateral lesions the dura is incised as shown in Figure 39. The edges of the dura are retracted and sutured into place so maximal exposure is obtained. If the lesion is in the midline or is bilateral a crossbow shaped incision is made (Figure 40). It is the policy at all times to obtain very adequate exposure of the lesion and not attempt to carry out removal through a limited opening of any type. The occipital sinus which runs down along the midline between the

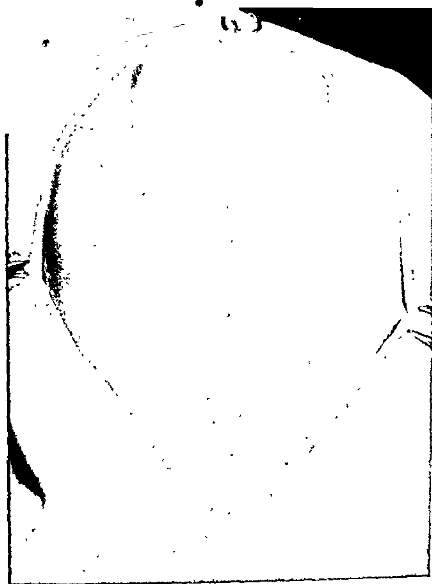
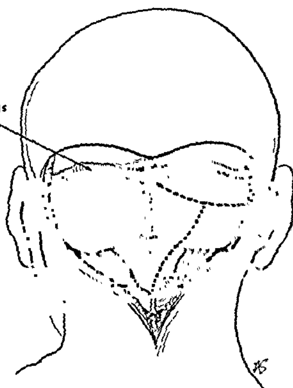


FIGURE 38 Method of draping patients for posterior fossa lesion. There is room to perform a ventricular tap, to extend the incision out to the mastoid region, and to expose the upper cervical cord.



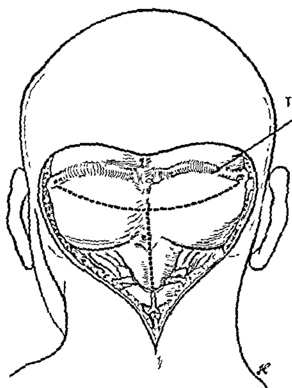


FIGURE 39 *(Top)* Incision (interrupted line) of dura used for unilateral cerebellar lesions The edges of the dura are then retracted so that the hemisphere and the cisterna magna areas are exposed.

FIGURE 40. *(Bottom)* Incision (interrupted line) of dura used for midline and bilateral cerebellar lesions. The dura is then retracted to expose the cerebellum. Usually the bone over the transverse sinus is not removed to the extent shown in this figure. It is usually removed so that only the lower part of the sinus is uncovered.

cerebellar hemispheres is either clipped with silver clips or ligated with a suture. Utmost caution should be used to insure total closure of the sinus. It must be kept in mind that this sinus, if the patient is in an upright position, can suck air permitting an air embolus if it is not ligated immediately after or before it is severed. Since a negative venous pressure may be present, bleeding from an open sinus or vein may not occur. It is emphasized that during any operative procedure in which the patient is in the upright position it is wise to have the anesthesiologist compress the rebreathing bag to produce an increase in intrathoracic pressure so that venous channels that might be opened will bleed and become visible. This compression of the bag is comparable to the Valsalva maneuver and obviously is not carried to the extent that there will be any herniation of the cerebellar tissue after opening of the dura. If there is evidence of increased pressure in the posterior fossa the dura is never opened until after the lateral ventricle is cannulated. This is accomplished by making a burr hole in the position usually used for ventriculography and inserting a No. 8 French urethral catheter as described earlier in the chapter for ventriculostomy. After the dura over the posterior fossa is decompressed the dura is opened. Occasionally, in spite of drainage of one of the lateral ventricles, the dura overlying the posterior fossa is very tense. This invariably infers the presence of a large underlying tumor. When this situation exists a small opening should be made in the dura overlying the cerebellar hemisphere that seems to be most tense and a ventricular needle passed into the cerebellar tissue. A cyst is usually encountered and when the cyst is drained the dura becomes sufficiently decompressed to permit opening If on the other hand a solid tumor is encountered it is usually possible to aspirate sufficient amount of this tumor to decompress the hemisphere. If it is not possible to aspirate part of the tumor the only recourse is to open the dura a small amount and to aspirate the cerebellar tissue overlying the tumor. When the tumor is encountered, it is removed by suction. The dura is opened in increasing extent as the pressure is released. Care must be exercised to prevent herniation of the cerebellar tissue or actual movement of the content of the posterior fossa, either of which might permit distortion of the brain stem or occlusion of vessels entering the brain stem.

If the ventriculostomy accomplishes an adequate decompression, the dura is opened in a T-shaped manner, carrying the lower end of the T down over the upper cervical cord. If there is a tumor out in one hemisphere the folia usually are widened and slightly whiter in color than normal. The tonsillar portion of the hemisphere often is herniated down through the foramen magnum. A ventricular needle should be inserted into the hemisphere. Either a cyst will be encountered, confirming the presence

of a neoplasm, or the neoplasm may be solid and a biopsy is necessary for confirmation. If the cerebellar hemispheres appear normal, although separated with a prominent bulging vermis, most probably the tumor is primarily in the midline. At times a bit of the tumor can be seen to project through the foramen of Magendie. When the tumor lies entirely within the fourth ventricle or high up in the vermis or cerebellar hemisphere the only deformity may be a slight herniation downward of the tonsillar portion of the hemisphere. Invariably the tumor is located on the side that the tonsil is herniated the most. If there is equal herniation the side of the lesion is indefinite and ventricular needles must be inserted into the hemisphere. If no tumor is found the fourth ventricle must be inspected. Occasionally it is necessary to make an incision in the lower part of the vermis to do so. If still no tumor is found one must look laterally around the cerebellar hemispheres, up into the cerebellopontine angle, or forward into the pons and medulla. One should never assume a negative posterior fossa exploration until all these procedures have been carried out. Perhaps the one exception to this is the circumstance in which there is strong suspicion the tumor is in the midbrain or third ventricle rather than in the posterior fossa and it is intended to carry out a Torkildsen procedure.

There are many methods used to remove the tumor, most of them are very acceptable. Only a few specific items warrant mentioning. An effort should be made to employ electrocautery as little as possible whenever in the posterior fossa and especially when one gets in the neighborhood of the pons or medulla. Extreme care should be taken to avoid damage to the posterior inferior cerebellar artery. This can be easily traumatized by too vigorous retraction, especially in children because of its more lateral position. With the patient in a sitting-up position one should get above the normal tissue. This prevents the necessity of any retraction of the tumor by the operator. In case the tumor is extremely large and heavy the operator must support the tumor rather than to permit it to fall away by itself. If there is only a thin shell of the cerebellar hemisphere left after removing the tumor, it is wise to pack the cavity with moist cotton until the dura is nearly closed. This prevents collapse of the hemisphere. An attempt is always made to close the dura after removal of a posterior fossa lesion because it is felt that this gives added support to the cerebellum, and it helps prevent development of a pseudomeningocele (bulging of the incision) or of cerebrospinal fluid fistulas. Following removal of the posterior fossa tumor a drain is usually placed into the tumor cavity and brought out through the lower end of the incision. The ventriculostomy tube is removed at the termination of the operation. The drain is removed 24 hours later and the delayed suture tied. Extremely meticulous care is used in closure of the muscles, the fascia, the subcutaneous tissue and of

the skin. There must be at least four layers to the closure to be certain a pseudomeningocele or fistula will not develop. When the patient is turned over from a face down position or is taken down from the sitting-up position the operator should hold the patient's head to prevent any appreciable flexion or hyperextension of the neck. The bandage that is applied is a rollar type either an Ace bandage or stockingette cut on the bias thereby permitting gentle compression of the line of incision. In patients in whom there is herniation of the cerebellar tonsils it is always imperative that an adequate decompression of the foramen magnum and of the upper cervical vertebrae is obtained in addition to an internal decompression by removal of the neoplasm. Some of the special techniques used with specific tumor types will be discussed with that tumor type.

Astrocytomas

Astrocytoma is the most frequent histological type of intracranial tumor in childhood. In this series there were seventy-three patients (26% of all intracranial tumors or 37% of the gliomas) with astrocytomas. In children they are usually located in the posterior fossa and in this series thirty-eight (52%) of them were in the cerebellum and ten (14%) were in the brain stem. In most series (Ford, Ingraham and Matson), the percentage of those in the cerebellum is even somewhat higher than this. As a general rule they are a very favorable type of tumor and almost any neurosurgeon considers himself and the patient fortunate to encounter this type lesion, rather than a medulloblastoma or ependymoma, when exploring the posterior fossa for an intracranial neoplasm.

The average age of occurrence of astrocytomas in the cerebellar region is from three to nine years with the peak incidence at about five to seven years of age. This is a somewhat higher age group than the medulloblastomas and some suggestion of which lesion is present can be obtained from this age differentiation. However, the difference is not sufficiently great to afford any certainty of diagnosis and dependency on this should never be complete. There is no sex dominance in astrocytomas occurring in the cerebellar region.

The histological characteristics of the astrocytomas have been described above in the discussion of the astrocytomas of the cerebrum. According to their gross appearance astrocytomas occurring in the cerebellar region are either of the cystic type or the noncystic type. The cystic type astrocytomas are characterized by having a very small, discrete, firm, well circumscribed nodule of tumor tissue, usually measuring 2 to 3 cm. in diameter with the mass of the lesion being made up of a single large cyst. This tumor nodule lies in the periphery of the cyst. The noncystic type astrocytomas may contain small or even fairly large loculated cystic areas

containing straw colored fluid, but they do not have a discrete mural nodule. Instead the gliomatous tissue extends nearly or entirely around the cystic cavity and the periphery of the tumor is diffusely infiltrative. Thus defined, there is about equal frequency between the cystic and noncystic types of cerebellar astrocytomas. The cystic type of astrocytoma is most usually found in the cerebellar hemispheres but occasionally they seem to arise in the vermis. When they arise in the vermis they invariably extend out into the hemisphere. The noncystic type may occur in either the vermis or the hemisphere and quite usually they occupy both positions.

Astrocytomas of the cerebellum are essentially midline lesions which develop asymmetrically through the tendency of the accompanying cyst or degenerative areas to expand into one hemisphere more than the other. However they do at times seem to arise entirely within one cerebellar hemisphere. The latter is especially true of the cystic type astrocytomas. Cushing (1931), Loisel, Bailey *et al.* and most other neurosurgeons believe that the cystic astrocytomas are the most benign and most favorable for surgical treatment of all intracranial tumors in childhood. In twenty-nine consecutive cases with two exceptions, Cushing believed he had removed all of the mural nodule from the cyst wall. Cerebellar astrocytomas, whether cystic or noncystic, are composed of fibrillary or protoplasmic elements in varying amounts. Cushing, Bailey *et al.*, and Penfield believe differentiation into fibrillary and protoplasmic types is of no clinical significance.

There are no symptoms or signs specifically diagnostic of a cerebellar lesion being an astrocytoma. Suggestive evidence of an astrocytoma is a history of long duration (years), presence of calcium in the lesion, evidence of erosion of the occipital bone, lack of signs of implants, and lack of cranial nerve palsies. Bailey *et al.* stated that when an astrocytoma was suspected preoperatively in their own series of brain tumors in children it was because of: (1) slowly developing symptoms, and (2) lateralization of cerebellar signs. The lateralizing signs most frequently observed were tremor, hypotonicity, dysmetria, and akiodokokinesis. According to Bailey *et al.* these indicate an involvement of the "hemisphere or *efferent* pathway and affect the arm as much or more than the leg." However, many other lesions occur in the cerebellar hemispheres. These include metastatic abscesses or abscesses that have arisen from direct extension from a mastoiditis and angioblastomas (usually in older children or young adults). Occasionally a lesion in the cerebellopontine angle will give predominantly cerebellar symptomatology. Consequently an accurate preoperative diagnosis is almost impossible to make—only a strong suspicion is reasonable.

Since it is practically impossible to be certain before operation that the lesion is an astrocytoma to the exclusion of other cerebellar lesions, preoperative irradiation to determine the type of tumor was suggested

by Cushing in 1931 and later Sosman. However, Bailey *et al.* and practically all other neurosurgeons at the present time believe this ill-advised due to the dangers of medullary compression and loss of vision. Direct surgical exposure and confirmation by biopsy is advised.

The most feasible incision for exposure of a cerebellar astrocytoma is *in the midline and extending from the external occipital protuberance to* the fourth cervical spinous process. This midline incision is considered preferable to the transverse or the "T" incision even for hemispheral lesions. The muscles can be stripped off the occipital bone and held to either side with a self retaining retractor. A trephine hole is made on each side of the midline and the bone removed with a rongeur, taking last the strip of *bone along the midline that overlays the occipital venous sinus.* The arch of the first and second cervical vertebrae are removed. If the dura overlying the cerebellum is tense, a ventriculostomy tube is placed into one of the lateral ventricles, and cerebrospinal fluid permitted to drain. Usually this decompresses the posterior fossa sufficiently to permit opening the dura in a "T-shaped" fashion with the lower end of the "T" going down into the cervical cord area. If on palpating the cerebellum through the dura it is obvious that the neoplasm is laterally placed it is wise to open the dura only on that side. If the tumor is not grossly visible an attempt should be made to look up through the foramen of Magendie into the fourth ventricle. If no lesion is encountered a needle is inserted into the hemisphere suspected of harboring the lesion. Either a cyst containing yellow colored fluid or a firm neoplasm should be felt. From there on the method of excision of tumor that is carried out depends much on the type of astrocytoma. If it is a cystic type, the cyst should be entered, the mural nodule recognized and excision of only the nodule accomplished. It is unnecessary to remove the cyst for the walls of the cyst are comprised of compressed cerebellar tissue not neoplasm. If the lesion is of the noncystic type a more extensive procedure is necessary. If possible the boundary of the neoplasm should be skirted and the tumor removed en-masse. It is believed wise to close the dura following removal of the tumor although this is not done routinely by all surgeons and is not an entirely necessary step. The reason the dura is closed in this clinic is to help prevent formation of a postoperative pseudomeningocele, although inadequate closure of the nuchal muscles is undoubtedly a more important contributing factor in the formation of a pseudomeningocele. It is felt a dural closure gives some added support especially if it is difficult to obtain good muscle approximation. In case a pseudomeningocele does occur it is best to aspirate the fluid and apply a compression bandage. This added support invariably is followed by obliteration of the sac.

The prognosis in cerebellar astrocytomas of the cystic type is immeasur-

ably better than for the noncystic type. This is true for immediate postoperative mortality, length of postoperative survival, and for freedom of symptoms following surgery. The reason for this difference is that the cystic astrocytomas are circumscribed, well localized lesions that do not infiltrate into an appreciable portion of the cerebellar tissue. Cerebellar and even brain stem dysfunction in a patient with a cystic astrocytoma is due to pressure by the tumor on the contiguous structures and not by actual tumor infiltration. When this pressure is alleviated and the small mural nodule removed, not only is function returned but also there seems to be no recurrence of the lesion. The prognosis in the noncystic type is not as favorable because this type infiltrates to such an extent that total removal is infrequent. However, these are slowly growing tumors and fairly long survival (5 to 8 years) can be anticipated if the neoplasm hasn't extended into the pons. If only subtotal removal is possible, a course of deep roentgen therapy is advised. It is appreciated, however, that these tumors are not particularly radiosensitive and this form of therapy doesn't add a great deal as far as their prognosis is concerned. If it is decided to radiate the patient, the deep therapy should be begun within five to ten days following the operative procedure. The radiation is given through three portals, one directly to the midline of the posterior fossa and then one laterally from each side of the posterior fossa. A total tumor dose of about 3,000 to 4,000 roentgens is administered over a period of twenty to twenty-five treatments.

If a child with a cerebellar astrocytoma that has been subtotally removed has a recurrence the question arises whether or not re-operation is ever advisable. Perhaps this decision is best left up to the individual surgeon; it is felt in this clinic that if the child's general condition is fairly good and the parents fully understand the increased risk of a second operation, it is wise to attempt further removal of the neoplasm. On a few occasions it has been found at the time of the second operative procedure, that the neoplasm seems to be more circumscribed and better delineated than at the first procedure and the surgeon has felt gratified after re-operation. It is emphasized, however, that total removal is *seldom accomplished unless* this can be done at the first procedure. This, of course, is true of all tumor surgery. In contradistinction to many other types of tumors in the cerebellum, the astrocytomas do not tend to implant or seed throughout the neural axis.

Medulloblastomas

Medulloblastomas comprise about 20% of all gliomas in children. Most authors have reported this tumor to be either the most frequent or the second most frequent intracranial neoplasm in children. In this series it was the third most frequent type being exceeded only by the astrocytomas

and the ependymomas. The usual age for occurrence is from the third to the sixth year of life but they may occur at any time during childhood. In the series of sixty-nine patients under twelve years of age with medulloblastomas reported by Ingraham and Matson, the favorite age of occurrence was three years. In this series of twenty-nine cases the average age was a few years higher, with the range extending from the second year up to the fourteenth year. There is a very definite sex predilection insofar as medulloblastomas occur predominantly in males. Cushing (1927) stated that they were three times more frequent in males than in females and Bailey (1930) found ten of thirteen cases to be in boys. It can be firmly stated that a lesion in the posterior fossa, especially midline, occurring in a male close to the age of five years has a much greater chance of being a medulloblastoma than any other type of lesion.

In an excellent review of the development of medulloblastomas Raaf and Kernohan concluded that medulloblastomas arise frequently if not exclusively from the cell rests which occur in the region originally occupied by the dermal bud of the external granular layer of the cerebellum in the posterior tip of the posterior medullary velum. There is certainly a close resemblance of the medulloblasts, which are the basic cell of the medullo-

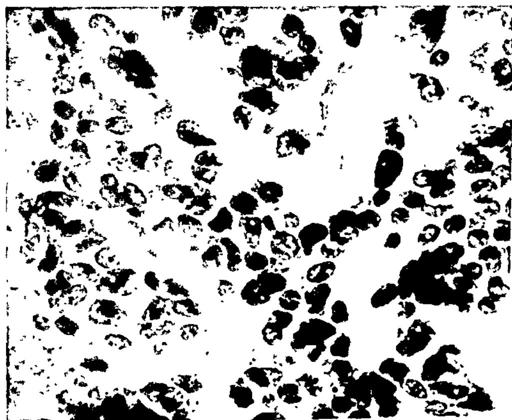


FIGURE 41. Photomicrograph of medulloblastoma. The predominant cell (medulloblast) is small, pear shaped, and contains a deeply staining nucleus. Spongioblasts and neuroblasts are also frequently seen in these tumors.



FIGURE 42. Photomicrograph of medulloblastoma showing diffuse spread of the tumor in the subarachnoid spaces over the cerebellar hemispheres. Note the resemblance between the medulloblast cell and the cells of the external granular layer of the cerebellum.

blastoma, to the granular cells in the cerebellum. This was first shown by Stevenson and Echlin. On gross examination these tumors are sharply outlined granular lesions that appear to arise from the posterior medullary velum. They are gray, friable, vascular, and seldom cystic. The predominant cell is the medulloblast which appears small and pear-shaped, and contains a round, deeply staining nucleus surrounded by scant cytoplasm. The medulloblast is a bipolar cell capable of differentiating into either spongioblasts or neuroblasts. The cells have a tendency to form pseudo rosettes, i.e., appear as a ball formation (Bailey 1930) or balloons (Kernohan 1938). In some areas the cells may be larger, the nuclei may appear more vesicular and may contain a nucleolus so that the cells resemble neuroblasts. Other areas may contain cells typical of spongioblasts and still others immature neuroglial cells. Usually spongioblasts wil predominate over the neuroblasts but occasionally the tumor will be composed almost exclusively of neuroblasts. There is very little connective tissue stroma and that present consists mainly of rather fine reticular fibers. Degenerative changes are rare. The blood vessels may show some hyaline degeneration. In 1925

Bailey and Cushing reviewed these tumors and recognized that both neuroblasts and spongioblasts were present and that the majority of the cells were similar to the indifferent cells of the cerebellum (Schaper). They suggested the name medulloblastoma.

Medulloblastomas like astrocytomas are found in the midcerebellar region of children but differ from the astrocytomas in being highly malignant, rapidly growing lesions that are prone to innoculate the cerebrospinal fluid. A congenital origin of these tumors is assumed because the cells are similar to the granular cells of the cerebellum, the location in the posterior medullary velum region, the occurrence in early life, report of their presence in new-born children and in identical twins.

It is extremely difficult to make a correct preoperative diagnosis of a medulloblastoma but it should be expected in patients three to eight years of age especially males who develop rapidly the signs and symptoms of a midline cerebellar tumor. Evidence of increased intracranial pressure (headache, vomiting, listlessness) was the most frequent early indication of this tumor in the University of Minnesota series; Lereboullet found intermittent vomiting was the first symptom in 60% of his cases. Ataxia usually occurs later. Since medulloblastomas are essentially midline lesions the ataxia is of a trunkal type. The patient tends to walk with a wide base, the Romberg test is positive and there is as much trouble when the child tries to sit up as there is when he tries to walk. Incoordination of arm and hand movements occurs late in the development of the symptoms or not at all. Calcification in sufficient amount to be visible in roentgenograms is rarely present although it does occur (Cushing 1923). The average duration of preoperative symptoms is six months and the postoperative survival period is about twelve months. Cushing reported one child who lived for five years but this is unusual. Bailey *et al.* have stated that the disease runs its course if left untreated in a year or less but as stated above there have been reports of much longer survival (Smith and Fincher). That medulloblastomas are radiosensitive is accepted but no case has been demonstrated to have been completely destroyed by this method of therapy. Frazier *et al.* stated in 1937 that medulloblastic cells developed a tolerance to irradiation so that deep roentgen therapy becomes decreasingly effective.

At the time of operation an apparently well circumscribed reddish friable tumor is visualized filling the fourth ventricle. However, more detailed examination reveals it to be infiltrative. In addition to filling the fourth ventricle the tumor may extend up into the aqueduct and out laterally through the lateral apertures of Key and Retzius. The tumor is friable and it is this friability that partly accounts for the tendency to spread via the subarachnoid space and through the ventricles to remote areas of the central nervous system. The ependymal lining of the ventricular system or



FIGURE 43. Medulloblastoma in vermis of cerebellum. Although there is some compression of the fourth ventricle the tumor is primarily in the vermis and does not fill the ventricle to the extent usually seen with an ependymoma.

the subarachnoid spaces around the brain and spinal cord may become studded with nodules of tumor tissue. Such implanted nodules may grow onto the cranial or spinal nerves and may give rise to root pains simulating intra-abdominal disease (Abbott and Kernohan). They may even produce signs of cord compression. As a result of spread over the cortex paralysis of cortical origin and epileptic seizures may occur. It is believed the proper form of therapy is to excise surgically all the accessible tumor. Beginning about the fifth postoperative day a course of deep roentgen therapy should be administered to the entire cerebrospinal axis. If the child is too restless, sedation can be given during the early period of radiation. To radiate the posterior fossa a total tumor dosage of 3,000 to 4,000 roentgens is advised; three portals should be used. Following this, 3,000 roentgens can be evenly distributed over the cerebrospinal axis. Radiation of the entire axis is not always done at this time because of the rather large total dosage of radiation necessary and because it is felt the radiation might more wisely be given when there is some evidence of implants or local recurrence. It is felt radiation therapy is palliative therapy, not curative. Because of the response of this tumor to deep roentgen therapy, and it is probably the most responsive of all intracranial tumors, it has been suggested that when

a midline cerebellar lesion is suspected, a course of deep roentgen therapy be administered prior to any surgical intervention. If the lesion is a medulloblastoma the tumor will shrink and symptoms may be alleviated but if no response to roentgen therapy is obtained a suboccipital craniectomy should be done because the lesion is more likely to be a sarcoma or astrocytoma. However, with this form of therapy completely remedial lesions such as cystic astrocytomas can be overlooked and the patient may go on to develop irreversible neurological signs or may even expire. As emphasized in the discussion on astrocytomas, it is believed this technique should be condemned. Another form of therapy used in patients suspected of harboring a medulloblastoma is to do a suboccipital craniectomy and biopsy the tumor. If on rapid section it proves to be a medulloblastoma the incision is closed and deep roentgen therapy administered. It is felt that this will reduce the chances of implantation that might occur with more radical excision.

However, in this clinic surgical manipulation of the tumor has not seemed to increase the frequency of the implants. Also it is felt that biopsy alone rather than resection of large amounts of the tumor does not afford sufficient decompression to be certain that complete obstruction to the flow of cerebrospinal fluid will not occur during the time deep roentgen therapy is given. It must be remembered that the usual reason these children come to the physician is because of signs of increased intracranial pressure which in turn is due to obstruction of the flow of cerebrospinal fluid. The tumors invariably swell when deep roentgen therapy is given so unless a very adequate decompression is obtained an obstruction will result. It is believed that rather extensive excision of the tumor tissue prevents or greatly decreases the chances of this block occurring, that there is a better chance for deep roentgen therapy to affect the remaining neoplastic tissue, and that the surgical mortality is not increased but rather diminished by this more extensive resection.

Even with all these various forms of therapy the ultimate prognosis for children with medulloblastomas is not good. The average survival time in our experience has been slightly over one year with some of the children going two or three years. This latter, however, is highly unusual and may mean that the tumor is more sarcomatous than medulloblastic.

Sarcomas

Sarcomas of the brain are rare lesions most frequently occurring in the midline region in the cerebellum of children. Occasionally they occur in the cerebellar hemispheres and even more rarely in the cerebrum. They were first described by Bailey 1929. In 1939, Bailey *et al.* reported five such lesions occurring in their series of one hundred brain tumors in

children. They are difficult to differentiate both clinically and by histological examination, from the medulloblastomas. The sarcomas do not show the predilection for males that the medulloblastomas do and in addition occur with about equal frequency during each year of childhood. There were four sarcomas in this series of 273 tumors.

The origin of these sarcomas (which do not arise from the dura as do

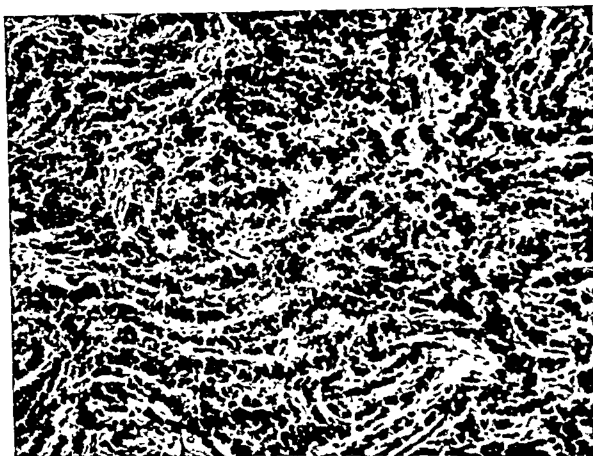


FIGURE 44. Photomicrograph of a sarcoma located in the vermis of the cerebellum. An hematoxylin and eosin stain was used. Note the similarity in histological appearance to a medulloblastoma.

the fibrosarcomas and malignant meningiomas) has not been definitely determined. In 1928, Schaltenbrand and Bailey while studying the perivascular tissue in tumors, especially medulloblastomas, noted that the connective tissue in some was very intimately mixed with the tumor tissue and in 1929 Nishii observed that many of the medulloblastomas contained fibrous tissue beyond that consistent with the usual medulloblastoma and he believed these were sarcomas. Bailey *et al.* found some were less responsive to deep roentgen therapy and had a different histological appearance than the average medulloblastoma. After applying various stains they concluded these less responsive tumors contained more reticular

fibers and were, in reality, not medulloblastomas but sarcomas. In 1940, Hsü reviewed the literature and concluded from this and his own experience that these sarcomas begin as true peritheliomatous growths of connective tissue origin. He stated that they spread mainly along the perivascular and leptomeningeal spaces but that they may break out of them into the parenchyma of the brain and then spread diffusely without form-

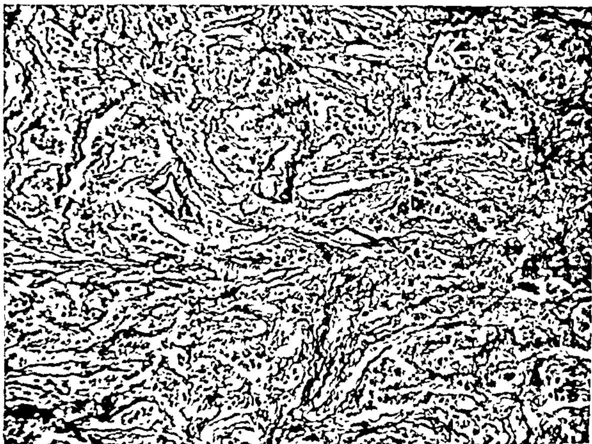


FIGURE 45. Photomicrograph of the tumor shown in Figure 44, only with a Perdrau stain applied. Note the heavy masses of reticulin fibers characteristic of a sarcoma of the cerebellum.

ing a capsule. These tumors are very cellular, contain numerous mitoses, and Perdrau or other reticulin impregnating stains are necessary for recognition.

At the time of operation sarcomas appear as dark red, poorly circumscribed, soft lesions that grossly resemble medulloblastomas. An attempt should be made to remove these tumors totally because they are much less responsive to deep roentgen therapy than are medulloblastomas. However, total removal is seldom if ever possible and the postoperative survival varies from one to two years. Sarcomas, like medulloblastomas may implant throughout the cerebrospinal axis.

Ependymomas

Ependymomas are the third most frequent neoplasm found in the posterior fossa in children. There were fifteen of these lesions found in the posterior fossa in this series of 273 brain tumors. The general histological characteristics of these lesions are described under the ependymomas in the cerebrum. Whereas any one of the various types of ependymomas may occur in the posterior fossa, in this series nine were of the cellular type and six of the epithelial type. According to Bailey, Buchanan and Bucy the tumors sometimes arise from the anterior or posterior medullary velum area but in this series and according to the experience *of most surgeons, ependymomas arise from the ependyma of the floor of the fourth ventricle in the region of the calamus scriptorius* and occasionally will rise out in the region of the apex of one of the lateral recesses of the fourth ventricle (Globus, Kuhlenbeck and Weller). In the process of growth these tumors expand into the fourth ventricle and eventually totally occlude the fourth ventricle. They also grow down from their origins in the ependyma or subependymal area into the

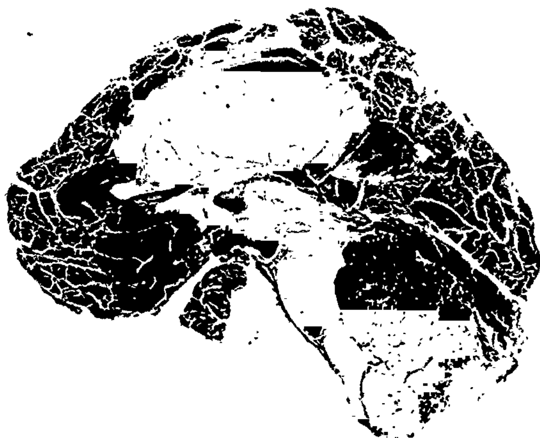


FIGURE 46. Papilloma that had occluded the fourth ventricle. There was obstruction to the flow of cerebrospinal fluid with secondary dilatation of the lateral ventricles, third ventricle, and aqueduct.

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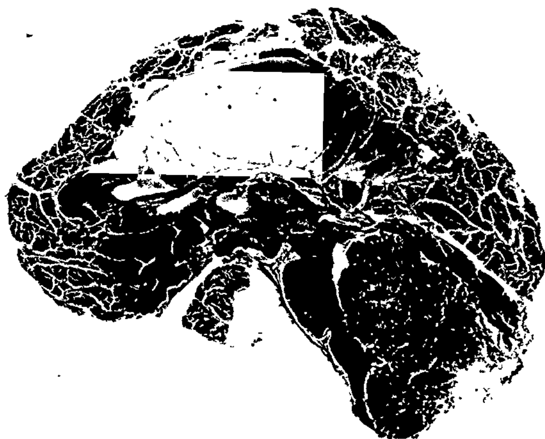


FIGURE 46. Papilloma that had occluded the fourth ventricle. There was obstruction to the flow of cerebrospinal fluid with secondary dilatation of the lateral ventricles, third ventricle, and aqueduct.

brain stem region. This infiltration into the brain stem region is the process that makes impossible the total removal of these tumors. Cushing and also Ford have emphasized that the blood supply to the tumor arises from the same vessels as those that supply the pons and medulla. Injury to these vessels results in embarrassment of the blood supply to the brain stem which usually is fatal. In 40% of the cases in this series the pons and medulla were directly infiltrated by the tumor tissue. Ependymomas seem to be well defined and encapsulated when first visualized but invariably after surgical removal has begun it becomes evident that they are infiltrative and total removal is impossible. A preoperative suspicion of the invasion of the brain stem can be obtained by the presence of deep tendon reflex abnormalities.

Ependymomas are usually histologically benign and slowly growing but due to their position are actually very malignant. The symptomatology is no different from other cerebellar tumors, but due to their position, signs of cerebrospinal fluid obstruction may occur prior to any evidence of cerebellar dysfunction. This is also true of long motor tract dysfunction. Evidence of long motor tract abnormalities occurred in this series in all but two of the patients with ependymomas (87%)



FIGURE 47. Cellular ependymoma arising from the floor of the fourth ventricle. This *is a malignant tumor that not only extends throughout the fourth ventricle but also* infiltrates deeply into the brain stem.

whereas only 50% of the patients with astrocytomas and 30% of those with medulloblastomas had evidence of long motor tract involvement.

Whereas, the clinical picture may consist only of increased intracranial pressure and long motor tract involvement, nuchal rigidity is also a frequent finding in patients with ependymomas. This symptom is caused by the tumor either growing down through the foramen of Magendie into the region of the foramen magnum or forcing the tonsillar portion of the cerebellar hemispheres down through the foramen magnum thereby fixing the meninges and spinal nerve roots so that any movement of the head causes traction of these structures with resulting pain. The stiff neck is, of course, a protective mechanism. Signs of focal cerebellar structure involvement occur later and this sequence of events is important in diagnosis (Bruns, Marburg 1920-21). Ependymomas may also directly invade or cause pressure on the pons and medulla so that early vomiting may be due to direct involvement of this region by the tumor rather than by generally increased intracranial pressure from obstruction to the flow of cerebrospinal fluid.

The range of age of patients with ependymomas in the infratentorial region in this series was from ten months to fourteen years. Average age was 6.8 years. There were eight males and seven females. The average duration of preoperative symptoms was 3.1 months.

The treatment of ependymomas arising in the fourth ventricle region is surgical removal of all accessible tumor tissue followed by deep roentgen therapy beginning on about the fifth postoperative day. The question may arise in a patient strongly suspected of harboring an ependymoma if it wouldn't be wise to administer deep roentgen therapy rather than to attempt any surgical removal, especially since the tumors are firmly attached to the floor and lateral walls of the ventricle. However, one can never be certain by any preoperative evidence that the lesion is not a surgically amenable tumor (astrocytoma) so that surgical exposure, at least for identifying purposes, is mandatory. It is believed in this clinic that rather than biopsy alone a rather radical excision of tumor tissue should be carried out in order to assure adequate decompression of the fourth ventricle. This is in contradistinction to the opinion expressed by Bailey that ependymomas located in the infratentorial region, when uncovered at operation, should be left alone since any manipulation may lead to death from injury to the cardiac and respiratory centers. However, the operative mortality in this clinic, using the rather extensive resection technique, has been less than 10% during the last eleven years and this is similar to the experience of Ingraham and Matson who reported one postoperative death in the seven children with fourth ventricle ependymomas operated upon in the four years prior to the publication of their book. Prior to the advent of the sitting-up position for posterior fossa



FIGURE 48. Epithelial ependymoma arising from the ependyma or subependymal tissue of the floor of the fourth venticle. There is extension out into the fourth ventricle but a lack of deformity and involvement of the vermis, as differentiated from the appearance of a medulloblastoma is apparent (Figure 43).

surgery and the many improvements in anesthesia the operative mortality was extremely high in this clinic and the opinion of Bailey as expressed above was concurred with but these improvements have been most helpful and the opinion concerning the resectability of ependymomas has changed to a more optimistic one.

Ependymomas have a tendency to spread via the cerebrospinal fluid and Van Wagenen has reported a papilloma arising from the choroid plexus that seeded itself throughout the ventricular and subarachnoid

spaces. It is believed this seeding may occur about as frequently without as with surgery. In our experience no patient has seemed to have had a dissemination immediately following surgery. In fact, it is not the policy to irradiate the entire cerebrospinal axis postoperatively in patients with ependymomas as it is in patients with medulloblastomas. Deep roentgen therapy is begun approximately five to six days postoperatively and 3,000 tissue roentgens are given through the usual three posterior fossa portals over a period of thirty days.

The prognosis in patients with ependymomas is not particularly good. The usual time of recurrence is two to four years following surgery, although some patients have survived five years prior to evidence of recurrence. When evidence of recurrence manifests itself a repeat course of deep roentgen therapy is advisable. In this clinic re-operation on patients with ependymomas has not been fruitful in the least.

Tumors in the Cerebellopontine Angle

Tumors arising in the cerebellopontine angle are unusual in children; there were only six in this series of 273 tumors (Figure 36). When they do occur they are most often one of three histological types *(neuroma, meningioma,* or *mixed tissue tumors).* These are all lesions occurring primarily in and limited to the angle. Other tumors, usually gliomas, may arise in the cerebellar hemispheres or brain stem and extend into the angle, however, they do not present symptomatology and signs primarily of an angle lesion and are therefore considered elsewhere.

In the differential diagnosis of lesions in the cerebellopontine angle one should consider almost exclusively a neuroma. This, of course, is especially true if loss of hearing can be demonstrated. A meningioma should be considered if there is minimal or no loss of acoustic or vestibular nerve function, if there is associated hyperostosis along the petrous ridge or adjoining occipital bone, or if there is an associated cerebral lesion that is considered meningiomatous. A mixed tissue tumor, usually epidermoid or dermoid, can seldom be differentiated preoperatively from other angle tumors although this tumor type should be suspected if the patient presents a history of recurrent episodes of aseptic meningitis. The very irritative content of an epidermoid cyst can seep out and produce such a meningitis. It really makes little difference as far as the surgical approach is concerned which of these three tumor types is present. The skin incision and craniectomy are carried out the same. Moreover, it should be possible under most circumstances to remove totally any one of these tumors. Conversely any one of the types can be sufficiently extensive or adherent to the brain stem to preclude total removal.

The characteristics of meningiomas and of mixed tissue tumors are given elsewhere; only detail of the neuromas will be presented here.

Intracranial Neuromas

Tumors originating in one of the cranial nerves are infrequent in children. When they do occur they usually are associated with generalized neurofibromatosis (von Recklinghausen's disease) and are therefore only one of the many stigmata of that disease. For example, in children acoustic neuromas are apt to be bilateral and associated with one or more of the following abnormalities: optic nerve gliomas, intracranial gliomas or meningiomas, café au lait type pigmentation, subcutaneous lipomas and peripheral neuromas, syndactylism, bone defects, hypertelorism, cleft palate, broad maxilla, flattening of the bridge of the nose, cataracts, or hemifacial atrophy.

Any one of the cranial nerves may be involved but the most frequent site of the solitary neuroma is the acoustic nerve. Next most frequently involved is the vagus, followed by the trigeminal and hypoglossal nerves. Several nerves may be involved, and the involvement varies from plexiform thickening to nodular enlargements. There is considerable controversy concerning the embryologic origin of the neuromas. It is accepted that they arise from nerve sheaths but there exists considerable difference of opinion as to whether the sheath is that of Schwann which is thought to be of ectodermal origin or the endoneural and perineural sheaths which are of mesodermal origin. In all these tumors, investigators have found considerable fibrinogin and collagen and the controversy has been whether the neurolemmal sheath of Schwann whose cells are ectodermal in origin and analogous to glial cells could form this fibrinogin and collagen or whether it could be formed only by sheaths derived from connective tissue such as the endoneurium and perineurium. Murray and Stout, in 1940, used the method of tissue culture to determine the basic type cell in these tumors and concluded that it was possible to differentiate Schwannian cells from fibroblasts by this method. They compared the Schwann cell outgrowth of tissue cultures of normal and grafted nerves with that of tumors. They concluded that the growth was similar, that these cells form collagen, and that these tumors are of Schwannian origin. However, Tarlov in the same year (1940) used staining techniques by which Schwann cells were impregnated but fibroblasts were not. Since he was unable to impregnate any of the cells in those tumors he concluded they were not of Schwannian origin but rather of fibroblastic origin. Masson, in 1923, reported that these tumors arose from the sheath of Schwann and called them Schwannomas. In 1932, he reported on the development of experimental and spontaneous

Schwannomas and stated that the advocates of the connective tissue origin of these tumors do not realize that the collagen and fibroglial fibers can be laid down by neurolemmal (Schwann) cells and hence a connective tissue origin is unfounded.

In some clinics a differentiation is made between the solitary neuromas and the multiple type associated with von Recklinghausen's disease. In this clinic such differentiation is not made because it is felt unnecessary from a practical point of view. The usual neuroma is an encapsulated, discrete, round, smooth tumor that is yellow-brown to red on gross appearance. It is usually relatively avascular, firm and fibrous although occasionally softened degenerated areas are present. Histologically these tumors are composed of fusiform cells with elongated nuclei containing a coarse chromatin network. The cells are arranged in sheaths or bundles that run in various directions in an interlacing manner. The nuclei tend to be arranged in parallel rows (pallisading). This is a most characteristic finding but at the same time not constantly present. Collagenous fibrils run throughout the tumor. Numerous areas of myxomatous degeneration may occur. Some of these tumors have a loose reticular structure and contain large lipoid filled macrophages which gives the tissue a



FIGURE 49 Photomicrograph of a neurofibroma removed from the left acoustic nerve of an eight-year-old child The pallisading of the cells visible in this tumor is characteristic of neurofibromas.

yellow color. Malignant degeneration within the tumor may occur but usually neurofibromas are benign; neither invading the surrounding tissue nor destroying the nerve upon which it grows. At the time of surgery an attempt should always be made to remove all the grossly visible tumor unless the patient's general condition precludes such an extensive procedure. These are curable lesions. Fortunately there is a fairly characteristic progression of symptoms when the neuroma is on the acoustic nerve. The first symptom usually is decreased hearing. This may be overlooked because of normal hearing ability on the opposite side. Attention should be paid to the child turning his head or cocking his head to one side to improve his hearing. School teachers often are particularly sensitive to this change in a child. Even if there are bilateral acoustic neuromas one side invariably is involved more than the other. One side may be affected a year or more prior to the other. The hearing loss advances slowly over a period of years.

Involvement of the vestibular division of the eighth nerve produces vertigo which may occur paroxysmally as in Meniere's syndrome but usually there is a more mild, chronic change from normal. The ipsilateral side of the face may become numb and chronic conjunctival irritation may result from decreased corneal sensation. Further symptoms include ipsilateral ataxia from cerebellar and/or brain stem involvement, ipsilateral facial weakness of a peripheral or nerve type, double vision from sixth nerve involvement (since this usually is secondary to increased intracranial pressure it may be unilateral or bilateral), and rarely a true weakness of an arm or leg from corticospinal tract compression.

Signs produced by an acoustic neuroma are fairly self evident from the structures involved. Papilledema results from blockage of the flow of cerebrospinal fluid, either from compression of the aqueduct or obstruction at the incisura tentorii, usually the latter. Examination of the cerebrospinal fluid obtained by lumbar puncture reveals a high protein content, usually over 100 mgm. %. The unfortunate part of all this, as inferred above, is that these are removable neoplasms but in children the high frequency of bilateral involvement or the presence of associated developmental defects make the general prognosis poor.

Surgical removal of an acoustic neuroma in a child is carried out no differently from that in an adult. Perhaps the less massive nuchal muscles and thinner skull make it easier in a child. Total removal is the procedure of choice since it offers the only assurance of a cure. The incision of choice is an oblique, paramedian one which makes unnecessary the detachment of muscles from the nuchal line. Excision of a portion of the cerebellar hemisphere usually is not necessary for adequate exposure. Great care must be taken to avoid compression of the posterior inferior

cerebellar artery in retraction of the hemisphere. In a child this artery seems more laterally placed than in adults and hence can be easily caught by a retractor. Prolonged compression of the vessel may result in ischemia of the brain stem. On retraction of the hemisphere yellow arachnoidal loculations may be present although these are less frequent than in adults. If present these cystic areas are opened and better visualization

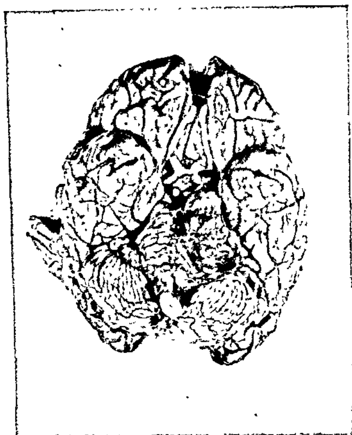


FIGURE 50. Acoustic neuroma that completely occluded left cerebellopontine angle in a 14-year-old boy. This child died suddenly; there had been no recognized symptoms of an intracranial lesion. The diagnosis was made at autopsy.

of the neoplasm obtained. The capsule of the tumor is opened and the intracapsular material sucked and curretted out. As the capsule is decompressed gentle traction on it will allow delivery of any portion extending up through the incisura. Too much or too rapid traction may result in trauma to the fifth nerve. Caution always is the byword! Not always is total removal possible because of close adherence of the capsule to the brain stem. Even though total removal is not anticipated the portion of the neoplasm that may extend through the incisura must be removed to assure opening of the channels for cerebrospinal fluid flow. Utmost care must be given to preservation of the seventh nerve. In adults total removal of an acoustic neuroma is tantamount to sacrificing

the aqueduct to one side is seen. Care must be taken, however, to differentiate this from the shift sometimes observed in patients with cerebellar hemisphere or cerebellopontine angle tumors.

By the above techniques a brain stem glioma can almost always be confirmed. However, if this is not possible, serious consideration must be given to surgical exploration as a means of verification. Obviously if at all possible this should be avoided, but occasionally a firm diagnosis cannot otherwise be established. There is considerable danger inherent in this type of exploration because one always is tempted to biopsy and/or remove sections of the tumor. Unfortunately it often is extremely difficult for a surgeon to be content with simple exploration and verification by gross appearance. Then, too, exploration is a useless procedure except for verification of the lesion since surgical removal of the tumor is out of the



FIGURE 54 Ventriculogram (brow down lateral view) of a patient with a brain stem glioma showing posterior displacement and irregular appearance of the aqueduct of Sylvius Even the fourth ventricle is compressed and more posteriorly placed than normal.

question. There seldom is indication for a Torkildsen procedure because these tumors usually do not obstruct the flow of cerebrospinal fluid.

Following verification of a brain stem glioma, the patient should receive deep roentgen therapy, not as a curative but as a palliative procedure. A tumor dosage of about 3,000 roentgens is used in this clinic. This therapy sometimes does not seem to alter the course of events but occasionally definite response has been observed and fairly long term survivals (four to six years) obtained.

Tumors of the Skull

There are a number of tumors that involve the skull. These include mixed tissue tumors (epidermoids, dermoids, and teratomas), osteomas, sarcomas, metastatic neoplasms from many sources, fibrous dysplasia, eosinophilic granulomas, Hand-Schüller-Christian's disease, and Letterer-Siwe's disease. Some of them obviously are not primarily skull lesions but only involve the skull secondarily. They come to the attention of the neurological surgeon because the presenting symptoms most generally are local tenderness or swelling, an asymmetrical shape of the skull or, occasionally, headache.

Epidermoids, Dermoids, and Teratomas of the Skull

Epidermoid and dermoid tumors arising in the bones of the calvarium develop from congenital inclusions of epithelial or dermal tissues. The inclusion material proliferates within the bone, the lesion increases in size, and there is a gradual absorption, due to pressure, of adjacent bone. This eventually leads to a well defined area of bone destruction. Since these lesions are fundamentally or primarily inclusion cysts they always are connected to the overlying skin, a connection that may be very apparent or may be visible only under excellent lighting conditions and after the scalp is shaved. Occasionally the overlying skin becomes infected—redness and swelling or even a draining sinus may develop.

The diagnosis can be confirmed radiographically. There is a smooth, usually rounded area of radiotranslucency surrounded by a sclerotic border. Oblique views may demonstrate whether or not the lesion has deformed the inner table or even has eroded through the inner table to involve the epidural space.

Histologically the cyst consists of a capsule of fibrous tissue and within the cyst cellular debris, granulomatous tissue, sebaceous material, and occasionally hair may be found. The cyst contents may be so infected that it appears to be a chronic abscess within the bone.

The most frequent sites of origin of epidermoids, dermoids, and teratomas are along the midline (sagittal and bregmatic sutures) or in the

lateral supra-orbital area at the junction of the frontal, sphenoid, and maxillary bones.

Treatment is surgical excision of the lesion. This should be done as soon as possible after recognition to avoid infection in the lesion which in turn may produce considerable deformity of the scalp or even extend intracranially. If the lesion is a teratoma (tridermal) there is reasonable assumption that malignant changes may occur in the tumor, again making prompt surgical excision advisable.

At the time of surgery one must be prepared to extend the dissection intradurally if such an extension is found. Ordinarily, however, it is necessary only to excise the dimple of skin overlying the lesion, go down through the outer table of the skull and dissect out the cyst wall, preferrably without spilling any of the contents. After the lesion is excised it



FIGURE 55 Roentgenogram showing an epidermoid (cholesteatoma) in the occipital bone of an 11-year-old boy. This lesion was excised seven years ago and the patient has had no evidence of recurrence to date.

is well to currette out the entire bed of the cyst to insure against leaving any remnant that may later provide a site for recurrence. Matson has advised raising some "curls" of bone from the adjacent outer table and reflecting these "curls" into the bone defect so that more complete healing of the bone will ensue. This seems like a very good procedure. Occasionally the bone defect will be so large that a more extensive cranioplasty is necessary.

There is another type of lesion that occurs in the mastoid region which is called an epidermoid or cholesteoma. These are inflammatory lesions that result from chronic mastoiditis with perforation of the tympanic membrane. The squamous epithelium of the external auditory canal grows through this perforation into the middle ear and mastoid antrum. The epithelium then proliferates, desquamates, and gradually over a period of time enlarges sufficiently to erode, from pressure, the mastoid. Radiographically these lesions appear as areas of radiotranslucency, beginning at the mastoid antrum and extending medially into the mastoid proper. The lesion is considered a problem for the otologist and is mentioned here to insure differentiation from the type of inclusion cyst mentioned earlier.

Osteoma

Osteomas are benign, slowly growing tumors which arise from either the internal or external tables of the skull. The majority arise from the outer table, near the periosteum, and project outward, deforming the overlying scalp. The only clinical evidence of such an osteoma is a firm, usually small, nontender, protuberance covered with normal skin. When an osteoma arises from the inner table it may project intracranially, deforming the dura but not invading it.

The histological appearance of an osteoma is characterized by a simple overgrowth of bone (osteoblastic activity). There is no evidence of malignancy. Roentgenograms reveal a dense, sclerotic-appearing area of bony tumor, non-invasive, and involving either the outer or inner table with no involvement of the diploë. Often the lesion is rather poorly circumscribed. There are no areas of bone destruction nor radiotranslucency.

Treatment consists of surgical excision of the lesion followed by cranioplasty. The indications for surgery include an attempt to improve cosmetically a deformed cranium and a removal of an inwardly projecting osteoma which may produce some neurological defect. The latter is extremely unusual.

Tumors Metastatic to the Calvarium

Metastatic tumors have been discussed earlier in this chapter on page 298. They are very uncommon in childhood. Rarely do they metastasize to the

skull but when they do the signs and symptoms are comparable to those seen in adults with metastatic tumors.

Fibrous Dysplasia

Fibrous dysplasia of bone has become established as a disease entity only in recent years. The etiology of the lesion is still unknown but most likely it is a congenital developmental anomaly in which there is abnormal activity of the specific bone-forming mesenchyme (Lichtenstein and Jaffe 1942, Neller). Other suggested etiological factors have been metabolic and endocrine disorders, a reactive response to injury, and hyperemia caused by anomalies of the sympathetic nervous system. It is very unlikely that any of the latter are causative.

The basic pathological change is an accumulation of fibrous connective tissue within the affected bone. The connective tissue may contain densely cellular and collagenous areas. Scattered throughout are islands of immature bone or cartilage. Occasionally imbedded in the connective tissues are areas of hemorrhage, small cysts, and bizarre giant cells. The lesion occurs commonly during childhood (age eight to twelve years) and becomes less active as maturity is reached. As the active phase of the condition is passed the fibrodysplastic tissue may become increasingly ossified. Obviously the roentgenological manifestations of the lesion are variable depending upon the extent and the phase of the lesion.

The condition may involve only one bone or many. Usually the bones of the skull show some evidence. Other manifestations of the condition include abnormal cutaneous pigmentation, premature sexual development, and hyperthyroidism. The term Albright's syndrome has been applied to those cases characterized by disseminated bone lesions, premature sexual development in females, and cutaneous pigmentation.

Because deformity of the skull may be the only or the most significant manifestation of the condition the neurosurgeon is called to see these patients. The presenting complaint usually is deformity of the skull, abnormally shaped orbits, diplopia, loss of vision, or unilateral exophthalmos. The lesion may extend into the facial bones with secondary facial asymmetry. Primary optic atrophy with decreased visual acuity may result from narrowing of the optic foramen or encroachment by the fibrodysplastic tissue on the orbital cavities.

The roentgenographic changes observed in fibrous dysplasia of the skull are not uniform. There may be translucent areas associated with widening of the diploë and thinning of the tables. The translucent areas have a dense margin. Perhaps more commonly there is extensive new bone formation which causes increased thickness and density of the bone. The



FIGURE 56. Roentgenogram of patient with fibrous dysplasia. There is extensive new bone formation with increased thickness and density of the bone. Interestingly this patient had very little cosmetic deformity or other symptoms of the condition.

bone of the skull, orbital ridges and adjacent facial bones are most commonly involved. The process is usually unilateral although it may extend across the midline.

Indications for surgical intervention in patients with fibrodysplasia include attempts to halt or improve a progressively developing visual disturbance and to improve the cosmetic deformity that occasionally occurs. The latter is very seldom necessary. However, in any patient in whom there is involvement of the sphenoid, frontal, or maxillary bones repeated studies of visual acuity, fields, and proptosis should be made. If there is progressive deforming exophthalmos or loss of visual acuity decompression of the orbit and optic nerve should be done. Through a transfrontal craniotomy the roof and lateral wall of the orbit should be removed and if there is appreciable involvement of the sphenoid bone the lesser wing should also be removed, continuing medially until the optic foramen is unroofed thereby decompressing the optic nerve. The results of surgery are very satisfactory in relieving the exophthalmus and in preventing further visual loss. Since the condition spontaneously ceases to progress when the period of active bone growth is completed there seldom is recurrence of symptoms.

Eosinophilic Granulomas

Eosinophilic granuloma was first reported as a clinical entity in 1940 by Otani and Ehrlich and by Lichtenstein and Jaffe. It is an unusual granulomatous lesion that occurs primarily during childhood or early adult life. The cause of the lesion is unknown; etiological agents varying from infections to trauma have been considered—none proved. It has been postulated (Green and Farber, Jaffe and Lichtenstein) that *Hand-Shüller-Christian's disease, Letterer-Siwe's disease* and eosinophilic granuloma are either the same or closely allied lesions. Swain and Williams have reported thrombocytopenic purpura accompanying eosinophilic granuloma as well as Letterer-Siwe's disease in infants and children. They should all probably be classified under *xanthomas.*



FIGURE 57 Roentgenogram of patient with Hand-Schuller-Christian's disease. There are multiple lesions. Clinically this patient had multiple soft, tender swollen areas in the skin over the calvarium.



FIGURE 58. Roentgenogram of en-bloc removal of eosinophilic granuloma. Obviously this type resection was too enthusiastic. Simple biopsy followed by deep roentgen therapy is a more advisable form of therapy.

The lesions may be solitary or multiple. The first symptom is usually a visible and palpable swelling over the cranial vault. The swelling is soft and has a doughy feeling. There may be local tenderness. Occasionally there is sufficient similarity to an inflammatory lesion to suggest a local abscess or even osteomyelitis. However, there is seldom if ever any peripheral evidence of infection (temperature elevation, leucocytosis, adenopathy). The diagnosis is confirmed by radiography. In Hand-Shüller-Christian's disease, there are multiple large irregular shaped areas of bone destruction (Mellbye). The lesions involve the orbit as well as the calvarium. In eosinophilic granulomas there are solitary rounded areas of bone destruction often with slightly sclerotic borders. These lesions can enlarge fairly rapidly. In Letterer-Siwe's disease there are multiple destructive lesions in the skull similar to those seen in Hand-Shüller-Christian's disease (Figure 57). It is difficult at times to differentiate eosinophilic granuloma from myeloma, metastatic neoplasms, dermoid cysts, or even localized osteomyelitis. It is

necessary to biopsy the lesion to confirm the diagnosis. The lesion is very radiosensitive so that after the confirmatory biopsy is obtained deep roentgen therapy should be instituted. The local lesion is curable but lesions may occur elsewhere in the skull or other bones of the body. If the disease is limited to the skull the prognosis for complete remission is good; if the lesions are more generalized a guarded prognosis for recovery is advised. Multiple lesions should be radiated. At the time of biopsy one is tempted to do an en-bloc type excision of the lesion (Figure 58), which has been done in this clinic on a number of occasions. In such instances the lesions have not recurred but the area of excised bone was of sufficient size that cranioplasty has been necessary. This can be avoided by simple biopsy of the lesion plus deep roentgen therapy. The latter form of therapy is now considered advisable.

At the time of biopsy these lesions appear to be red to purple in color and are soft and vascular. They can be readily curretted out. They may perforate both the inner and outer tables of the skull or may only bulge into the tables. The dura is never penetrated. Histological examination of eosinophilic granulomas reveals very cellular lesions composed mainly of round deeply staining eosinophils and histiocytes with very little intervening stroma. Large foam cells may be dispersed throughout the lesion. There may be large areas of necrosis.

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CHAPTER XIII

Tumors—Intraspinal

GEORGE AUSTIN

THE INCIDENCE of spinal neoplasms in childhood is low, however the process produces the same disastrous effects as in the adult if not recognized and properly treated.

As pediatrics and neurosurgery have grown and neuropathologic processes are no longer looked on as uniformly hopeless by many practitioners, diagnostic study of neurologic abnormalities is being more frequently carried out. For this reason more correct diagnoses are made of spinal tumors in children.

Frazier, in 1918, and Elsberg, in 1925, found only five spinal tumors in patients under twelve years of age in spite of their wide experience and profound interest in the spinal cord. During the past thirty years Stookey, Craig and Mitchell, Hamby, Svein *et al.* were all able to collect and study a far larger number of childhood spinal tumors. This points out the increased incidence of correct diagnoses. Ingraham and Matson, in 1954, relate their experience with sixty-three cord tumors in the Children's Medical Center in Boston in their excellent monograph on *Neurosurgery of Infancy and Childhood.*

In the series of 409 spinal cord tumors seen during the past twenty-five years at the University Hospital and the Graduate Hospital of the University of Pennsylvania, only thirty occurred in children up to the age of fourteen years. Figure 1 shows their relative frequency at the different age

INCIDENCE OF 409 SPINAL CORD TUMORS

Age	Number
0-9 Years	10
10-15 Years	20
16-19 Years	26
20-29 Years	54
30-39 Years	54
40-49 Years	84
50-59 Years	100
60-69 Years	53
70-79 Years	7

FIGURE 1. The incidence of spinal cord tumors according to age group.

groups. There seems to be no predilection as to the level, but rather, they occur in random positions over the entire vertebral column (Figure 2). Their frequently obscure mode of onset, often associated in onset of symptoms with trauma of one form or another, and the fact that they occur in a rapidly growing child, all make them extremely difficult to diagnose in

Position of Spinal Cord Tumors in Children	
Cervical	8
Thoracic	14
Lumbar	5
Sacral	3

Figure 2. The position of occurrence of thirty spinal cord tumors in children.

their early stages. The adaptability of the nervous system of a child further increases the difficulty of diagnosis. Reflex patterns have not become firmly developed and new conditioned reflexes are constantly being established. Because one is dealing with a growing individual whose muscle power and size is continuously changing, the neurologic deficit is frequently obscured as it occurs in the early stages of a tumor of the spinal cord. The rapidity of onset was one of the most predominant findings, so that it is unusual for the symptomatology to be present more than six months. Because children are continuously suffering from slight injuries during their play activity, it is often difficult to evaluate a temporary weakness in an extremity. The onset may begin with a fall from a bicycle, after turning a somersault, or following diving into the water. In 50% of the patients the clinical picture developed in three to six months, a third of the patients had a one to three months history, and in 10%, the symptoms were of less than one month duration.

Diagnosis

The diagnosis of a spinal cord tumor begins with the general practitioner who first sees the child. It is he who must be alert to the developing neurologic disease For if a child begins to develop progressive neurologic signs either in the motor, sensory or sphincter spheres, it is he who must recognize the progressive nature of the disturbance and call for help. In other words, it is a high index of suspicion of any progressive neurologic disease of the patient in which there is increasing disability of the somatic muscles, the subjective complaints of numbness or peculiar feelings of the body, or persistent and increasing evidence of urinary retention or incontinence, which should call for neurologic investigation.

In the presence of a spinal cord tumor he can frequently see with gross testing and observing the patient that segmental motor weakness is developing. The disturbance in motor power in the child who has learned

to walk manifests itself as a disturbance in gait. It should be the aim of anyone in medicine to try to diagnose progressive neurologic disease occurring, before there is a complete paraplegia. For as we shall see, it is in the paraplegic patient that the poorest results of surgery of spinal cord tumors is present. Any unexplained gait disturbance which seems to be getting worse is cause for neurologic evaluation.

Sensory changes early in the development of a spinal cord tumor are far more difficult to detect since numerous complaints of childhood are frequently casually dismissed. It is, however, important to grossly check the child with the disturbed gait or muscular weakness for a segmental distribution of any sensory loss that may be present. Cord tumors progressively produce an ascending sensory disturbance from the perianal region, the legs, the trunk, to the upper extremities. Any child with inability to perceive pin prick in the saddle area or on the legs should be sent for prompt evaluation of the neurologic status.

The child who presents as an infant with bladder dysfunction consisting of dribbling of urine where previously urinary control existed, should be examined to see if a distended overflow bladder is present. This should immediately alert the practitioner to the possibility of a cord bladder. Evaluation of such problems is a true emergency.

At times in lesions of the cauda equina there may be no motor or sensory loss. There may be no vesicle disturbance. The child merely complains of severe pain and muscle spasm in the spinal muscles and those of the hamstrings. This is particularly characteristic of the children harboring a tumor of the cauda equina. Therefore unexplained pain in the low back associated with pain and muscle spasm of the legs should again alert the practitioner to the possibility of progressive neurological disease and have him seek consultation for his patient.

It is well to remember that with tumors of the cauda equina, reflexes may be absent or unequal. Tumors of the actual cord substance produce exaggerated reflexes in the legs while normal reflexes are present in the upper extremities. In a cervical cord lesion all reflexes are exaggerated. Pathological reflexes accompany the exaggerated reflexes.

Given the situation in which the child is believed to have a spinal cord lesion, we pass on to the more refined diagnostic procedures which should be carried out by the neurological specialist who ultimately sees the child. At this point, motor power should be more carefully determined. Specific muscles should be tested and preferably examined, both by electromyography and chronaxie determination (see electrodiagnostic methods). Further evaluation of the bladder and rectal sphincters is particularly important. If there is any suggestion of bladder dysfunction, it is wisest to determine quantitatively by cystometrogram just how badly the bladder

is affected and whether or not it has become atonic to any degree. When bladder dysfunction is a problem, it is wisest to place a Foley catheter in the bladder and as soon as the diagnosis is made, begin treating the child by tidal drainage with daily irrigations of ½% citric acid solution. A slightly less physiological but alternative mode of treatment is to clamp the indwelling catheter and release it at four hour intervals.

Although pin prick will usually give a relatively accurate idea as to the sensory level produced by a mass lesion, it is only by careful testing of the sensation of touch that one can determine the exact level. In this fashion, testing with von Frey hairs, we have occasionally been able to reveal a level as much as six segments higher than the original sensory level revealed by testing with pin prick. This, of course, requires a cooperative child patient, and patience on the part of the examiner.

If a true paraparesis or paraplegia is already present before surgery, it is well to anticipate the child's needs at this time for improved venous circulation. An oscillating bed with a variable pressure mattress serves best to relieve stasis of the blood in the extremities and decrease the tendency for skin breakdown. For this reason, it seems best to place the patient on both an oscillating bed and variable pressure mattress as soon as the diagnosis of spinal cord neoplasm is made, if there is marked weakness of the extremities or any tendency toward deterioration of the skin.

The commonest preoperative objective finding is a sensory level to pin prick present in over half the patients. Pain, the most frequently encountered initial symptom, is found also in half the patients, including 10% with intramedullary tumors. Spinal shock is present in 20% and spasticity in 40% when first seen in the hospital. The relative frequency of spinal shock in this group attests to the extreme rapidity of the tumor growth in these patients. Whereas the tendon reflexes will usually be completely missing in cases of spinal shock, it is usual for the skin reflexes to persist to some degree so that one usually finds some slight plantar flexion of the toe

	Extra Dural				*Extra Medullary*				*Intra Medullary*		
Preoperative Findings	*C*	*T*	*L*	*S*	*C*	*T*	*L*	*S*	*C*	*T*	*L*
Spinal Shock	1	2			1					2	
Spasticity of Legs*	1	1			1	6			2	1	
Bladder or Bowel Dysfunction	2	4	2	1		2	3			1	
Pain as Initial Symptom		2	2	1	1	2	3	1	2	1	
Weakness as Initial Symptom	1	3			1	4			2	1	
Sensory Change as Initial Symptom					1					1	
Paraplegia (Total)	3	4	1			1	1		1	2	
Sensory Level to Pin Prick	2	4	2		1	4	1		2	2	

* Bilateral Babinski, increased tendon reflexes, ankle clonus, and increased resistance to passive movement

FIGURE 3. Frequency of occurence of preoperative neurologic findings in thirty spinal cord tumors in children. Findings classified according to position of tumor.

remaining present, in addition to positive cremasterics. However, these reflexes fatigue very rapidly in the state of spinal shock.

The following three case reports illustrate the points mentioned. The position of the tumor is different in each case and emphasizes the symptomatology which tends to be present as extradural, intradural, and intramedullary lesions develop.

Case 1. Intradural Extramedullary Tumor

A. Z., a thirteen year old white male, was admitted to the University Hospital in August, 1933, with a two months history. In the beginning of June it was noted that he had weakness of the left leg and foot with a tendency to catch his toe on the floor as he walked. This weakness gradually progressed until it was necessary to swing the left leg in a wide arc to prevent falling. Since the middle of July, he also had developed weakness of the right leg, and was barely able to walk without support on admission.

This patient did not have pain. He complained of some numbness over the chest going down over the legs. The weakness became so severe that eventually he came into the hospital to have something done about it. Neurological examination revealed a bilateral paraparesis much more severe in the left leg. There was diminution to all sensory modalities up to T7 bilaterally. There was bilateral spasticity of the lower legs with occasional involuntary flexor reflexes. Tendon reflexes were generally accentuated in both legs, and there was a bilateral Babinski reflex. A lumbar puncture was done which revealed a complete block to Queckenstedt. The CSF protein was 40 mgm. %. X-rays of the thoracic spine were entirely negative. Because of the progressive development of the case, it was felt that the boy had a spinal cord tumor in the high thoracic area. A laminectomy was done which revealed a very large neurofibroma at T4-T6 lying anterior and to the left of the spinal cord. The tumor was completely removed and the dura closed. The child made an uneventful postoperative recovery and was discharged September 30, 1933. When last heard from, in August, 1954, twenty-one years later, the patient was entirely asymptomatic and working full-time in a factory.

Case 2. Extradural Tumor

B. H., a thirteen year old white female, was admitted to the University Hospital in August, 1942, complaining of loss of bladder control. One and a half years previously, the patient had first noticed that a slight stiffness of the spine was apparent whenever she was stooping over for an object on the floor. Shortly after this, she commenced to complain of progressive rectal pain and aching of the right leg going down to the knee. Six months prior to admission she noted numbness to pain and touch in the saddle region extending out to the right buttock and right thigh. Also, at this time she suddenly became completely incontinent of her urine and feces. Several weeks earlier she felt that she had first noticed

her incontinence following several falls which had occurred while roller skating. At this time the family doctor advised her to stop skating. On admission, the neurological examination showed a limitation of spinal flexion. The right calf and foot were atrophied. There was a saddle type of anesthesia to touch, pain, and temperature which extended down the posterior aspects of the right leg to involve the foot and toes. The right patellar reflex was more active than the left. Weakness in mobility involved the entire right leg. She was completely incontinent of urine and feces. A lumbar puncture disclosed a negative Queckenstedt with spinal fluid protein of 40 mgm. %. X-ray examination of the spine showed marked erosion of the bodies and laminae of L5 and S1. At operation, a large extradural meningioma measuring 6-8 cm. was removed from the lumbo-sacral area with involvement of the bone due to pressure. The patient was last heard from in September, 1954, twelve years after operation, at which time she was carrying out a normal life with no neurological difficulties of any type. She was married and the mother of two children.

Case 3. Intramedullary Tumor

R. O., a twelve year old white boy, was admitted to the University Hospital in June, 1952. He had been complaining of abnormal weakness of the legs during the past month which began with numbness of the right foot. This was not disabling and the child went about his play and school work in the usual manner. A week before being admitted to the hospital the patient fell from his bicycle, and within twenty-four hours noted a numbness of the left foot. Shortly after this his mother observed that he walked with a slightly dragging gait and that he was having some difficulty in voiding. When admitted to the hospital the following day the patient had a flaccid paraplegia with a sensory level to T9 bilaterally. He appeared to be in a condition of spinal shock with loss of all tendon and skin reflexes with the exception of the cremasteric and anal reflex. Priapism was present and also some nuchal rigidity. X-rays of the spine were normal and a lumbar puncture showed an initial pressure of 140 mm. of water with clear and colorless spinal fluid and a negative Queckenstedt. A myelogram was carried out showing a fusiform dilatation of the opaque column extending upward at the level of the 9th thoracic interspace, but the interpretation was "indefinite." Twelve days later the myelogram was repeated and this time showed a definite change at the level of T9-T10. A laminectomy was then undertaken and a tumor was grossly removed by judicious section after incising the longitudinal and posterior columns. However, it was impossible to accomplish a complete extirpation. The tumor was diagnosed as an ependymoma. No motor function returned but there was some degree of sensory return. The patient was treated with x-ray therapy and survived the operation approximately eighteen months, eventually dying at home of tumor recurrence

DIAGNOSIS OF SPINAL CORD TUMORS IN CHILDREN

18—Myelography Required
11—Routine X-rays Diagnostic
17—(+) Queckenstedt
7—(−) Queckenstedt
6—Queckenstedt Not Recorded
13—Prot. $>$ 100 mgm. %; of Which 11 Had Queckenstedt

FIGURE 4. Shows the number of spinal cord tumors in children diagnosed by myelography, and the number that could be diagnosed by routine spinal x-rays. Also recorded are the results of the Queckenstedt test in these patients.

Specific Diagnostic Procedures

Once the clinical evaluation has suggested the presence of a spinal cord tumor, it is good judgement to proceed as rapidly as possible with definitive laboratory studies that will permit surgery at the earliest possible time.



FIGURE 5. A.W.· Shows erosion of the left side of the sacrum by a Giant Cell tumor.

Spinal cord tumors should be considered as acute surgical emergencies; for one is dealing with prolonged compression of neural tissue which, as we know, can only temporarily survive under conditions of impaired blood supply and actual damage by pressure. This is emphasized by the fact that one third of the patients in the Philadelphia group were already paraplegic before surgery. The chances of greatly improving these cases is slim indeed, hence the urgency to operate before the onset of any permanent cord damage.

Radiography

Roentgenograms of the spine should always be done first, for often one can make the diagnosis by this means alone. Not only can the presence or absence of tumor be determined this way, but often the level, and even

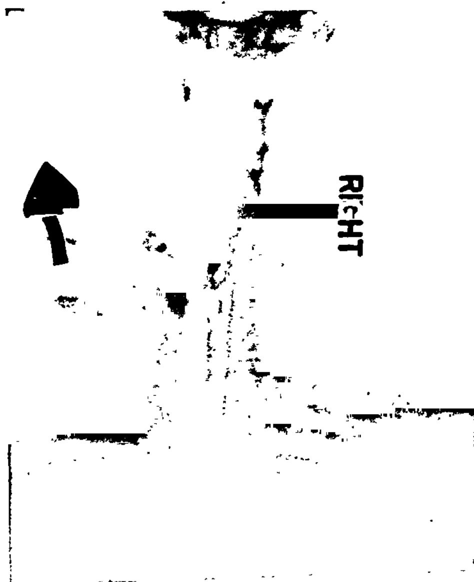


FIGURE 6a. P.K : Myelographic defect due to extradural tumor at T4.



FIGURE 6b. Extramedullary tumor at T2-T4 (meningioma), as seen at operation.

the type of tumor. In a series of thirty tumors, eleven were diagnosed by merely routine spine films (Figure 4). Of the eleven tumors diagnosed by plain x-rays of the spine, six were extradural, three extramedullary (intradural), and two were intramedullary. The changes usually seen consist of gross destruction of the bone, widening of the interpedicular space, or erosion of the pedicles of the spine. A tumor displaying marked

erosion of the sacrum is shown in Figure 5. Frequently it will be necessary to heavily sedate infants in order to maintain complete immobility during the taking of the films.

Lumbar Puncture

This is the second most valuable definitive procedure. Although considered a relatively harmless diagnostic study, it must be emphasized that in spinal cord neoplasms (especially at the cervical level) there will follow in a small percentage of cases a rapid onset of hemiplegia or quadriplegia within as little as four to six hours. For this reason, one should carry out



FIGURE 7a R.O. Myelographic defect due to intramedullary tumor at T10.



FIGURE 7b. Intramedullary ependymoma outlined by pantopaque® in Fig. 7a, as seen at operation

all steps possible with one lumbar puncture. Therefore, at the time of lumbar puncture 3 to 6 cc. of Pantopaque® should be available so it can be immediately inserted if myelography is indicated. Manometric readings are taken before any fluid is removed. Gentle bilateral jugular compression is then applied and a note made of the height to which the fluid rises in the manometer and the number of seconds required for it to rise. As a

rule, the spinal fluid pressure will tend to rise approximately three to four times the original pressure in less than five seconds, and fall over a similar period to its resting level. If it fails to do so the result is usually interpreted as either a positive or a partially positive Queckenstedt test, depending on the degree and rapidity of rise and fall. A specimen of spinal fluid is taken for serology, cells, and most important of all, the protein



FIGURE 7c. Intramedullary spongioblastoma polare, C7-T3, as seen at operation.



FIGURE 7d. Intramedullary ependymoma, C2-C6, *as seen* at operation.

determination. About 55% of patients had positive Queckenstedt tests. This, of course, immediately confirmed the presence of a spinal lesion with compression of the spinal subarachnoid space and block of normal flow (see Figure 4).

Elevation of the protein content, which occurred in about one third of the group of tumors, is further supportive evidence that normal spinal fluid circu-

lation is impaired. Usually, the degree of block will be somewhat in proportion to the protein elevation. When the cerebrospinal fluid clots in a test tube (Froin's syndrome), the protein is usually over 1,000 mgm. and may be taken as a presumptive evidence of a complete block.

Myelography

If the Queckenstedt has been positive to any degree, 3 to 6 cc. of Pantopaque® should be instilled into the spinal canal at this time to confirm the



FIGURE 8 J R. Pantopaque® injected cisternally to show the myelographic defect at the upper pole of the tumor (intradural neurofibroma).



FIGURE 9. Giant Cell tumor of bone at T6-T7, as seen at operation.

actual level of the lesion. The child is immediately taken to the x-ray department, placed on the table and, in less than a half hour from the time of the initial lumbar puncture, one is aware of the completeness of the block and the position of the lesion. Generally, moderate sedation in the form of seconal, demerol, or atropine is quite sufficient anesthesia for the lumbar puncture. However, under the age of ten years general anes-

thesia may be used if the child is very active, but from ten to fifteen years preliminary sedation is adequate.

In infants, 3 cc. of contrast media is sufficient for the diagnosis. After the age of two years, the quantity of contrast media may be varied from 3 to 6 cc., depending on the size of the child. Children over ten years of age are treated as adults in this respect and receive 6 cc. for thoracic and lumbar myelograms, and 9 cc. if the cervical canal is to be visualized. Following fluoroscopy and the taking of spot films to show any block or deviation in the radiopaque dye, the oil is removed by gently aspirating with a 2 cc. syringe. Figure 6a shows clearly the defect seen in the upper thoracic region by an extradural tumor at T4. In Figure 7a, a well defined intramedullary tumor has been outlined by Pantopaque® which is blocked at T10, and then flows minimally around the tumor to outline the entire contour of the swollen cord. Occasionally, where there is a complete block by Queckenstedt with no spinal fluid flow, it becomes necessary or expedient to perform a cisternal puncture followed by the introduction of oil. In this fashion, as the oil passes down the canal, it will be blocked and outline the upper level of the tumor. Figure 8 shows a case of this type where the radiopaque oil has outlined the upper level of a tumor at T1. In this type of case the oil cannot be removed until surgery, and it is advisable to have the patient's head elevated to prevent Pantopaque® from passing into the basal cisterns.

Pathology

It is interesting to note that in 30 patients, there were 14 different tumor types. Figure 10 shows the different types of tumor with regard to their location. It is difficult to draw any definite conclusions from a table of this type, although it should be noted that the intramedullary tumors are all malignant in character, whereas the extradural and extramedullary tumors

PATHOLOGY OF SPINAL CORD TUMORS IN CHILDREN

Extra Dural (12)		*Intra Dural (Extramedullary) (12)*		*Intra Medullary (6)*	
Giant Cell Tumor	(3)	Meningioma	(5)	Astrocytoma	(3)
Sarcoma	(2)	Neurofibroma	(2)	Ependymoma	(2)
Abscess	(2)	Neuroblastoma	(1)	Spongioblastoma	(1)
Neurofibroma	(1)	Ependymoma	(1)		
Neuroblastoma	(1)	Cystic Tumor	(1)		
Tuberculoma	(1)	Vascular Malformation	(1)		
Cyst	(1)	Neuroepithelioma	(1)		
Lymphoblastoma	(1)				

FIGURE 10. The various tumor types which occurred in thirty spinal cord tumors of children Classified according to position.

are predominantly benign. The same tumor type, such as ependymoma, may occur both intramedullary or extramedullary. Ependymomas that are extramedullary occur in the sacral region involving the cauda equina. A similar situation is noted in the neurofibromas where two occurred in an extramedullary position, and one occurred in an extradural position.

The reports from other clinics indicate that there is considerable variation in the tumor type encountered. Although meningioma was the commonest tumor encountered in the University of Pennsylvania group, teratoma comprised the largest cell type (20%) in the series reported by Ingraham and Matson, while meningioma represented only 3% of their group. They encountered dermoid cysts (15%) and astrocytomas (15%) next in order of frequency. Svien *et al.*, reported that neurofibroma (20%) was the commonest type of tumor and lipomas (15%) were next in order of frequency.

It is interesting that of the tumors we have reported there were neither dermoid cysts nor lipomas present. Whether this is due to the difference in pathological diagnostic methods, geographical location, or the size of the different series is difficult to determine. However, it seems most likely that the representative series are not of sufficient size to be statistically significant in this matter.

Surgical Approach

Surgical exposure and removal of a spinal cord tumor is performed by laminectomy. The technical details of the procedure are similar to that of the adult in principle although because of the diminutive size of the patient and his delicate tissues certain problems are present.

The first of these is fixation of the patient on the operating table in such a manner that surgery can be easily performed. With adequate precautions for fluid administration and control of body temperature, the child may be secured to a board or to the operating table with adhesive tape so that there will be a lack of motion. Attention should be directed to supporting the child under the pelvis and under the arms so that there is no compression of the abdomen or thorax. This helps materially to allow the venous channels of the epidural space to remain collapsed and prevent excessive venous bleeding.

Draping of the child should be arranged over the instrument table or supported by infusion poles so that when the drapes become soiled with irrigating fluid or blood they will not produce excessive weight on the patient's body.

A drop of dye injected into the subcutaneous tissues at the level of the lesion at the time of myelography will simplify localization of the incision so that valuable time will not be lost in finding the correct level of the

lesion. An adequate incision above and below the level of the lesion will permit sufficient exposure and lessen chances of trauma to the cord by virtue of manipulations through a cramped operative site.

With the level of the lesion delineated by myelography, it is well to begin two vertebrae distant from the lesion to again permit adequate bony exposure.

The skin and subcutaneous incisions are made rapidly. After coagulation of the bleeding points, the fascial attachments are severed from the spinous processes and interspinous ligaments as close to the midline as possible. Bleeding points are picked up and cauterized. The musculature is stripped from the spinous process and lamina of each succeeding vertebra on one side using a sharp small knife to sever the muscular attachments dissecting subperiostially with an appropriate elevator. A long gauze sponge is rapidly packed into place as soon as the muscles are loosened. When the muscles are thus packed back from the spinous processes and laminae on both sides, the wound is allowed to rest for a minute while the blood loss is assessed and replaced. This also allows for clotting to occur in the small vessels thus transected.

The packs are then removed rapidly and small self-retaining retractors are inserted to retract the muscles and continue the hemostasis.

The size of the patient then determines the instruments used. The spinous processes should be rapidly removed and the rough bony surfaces waxed. It is then possible to begin bony removal two vertebrae below the lesion with small rongeurs or Kerrison type punches. Bone is removed, rapidly working up to the lesion by cutting out a middle channel of bone and ligaments. Once the mass lesion is encountered one must proceed with caution for fear of adding trauma to the pathologic cord.

The situation of the tumor relative to the dura should be estimated. If the tumor is extradural, then a line of cleavage between tumor and cord can be discerned so that bony removal may proceed rapidly. If the dura is tense from intradural tumor, the utmost of care should be exerted to prevent any compression of it as the bone is removed. When the upper pole of the tumor has been identified, bony removal should proceed rapidly again until two vertebra have been passed. This will give adequate exposure above the tumor for manipulation.

Certainly, care and gentleness in handling the cord will produce a smoother postoperative convalescence. If the tumor is extradural, it is usually well to avoid opening the dura at all. This procedure will only serve to permit extradural cells to penetrate within the dura and perhaps cause additional and more severe compression. Similarly, if the tumor is intradural but extramedullary, it is well to preserve as much of the arachnoid covering as possible in order to avoid damage to the small blood

vessels running in the arachnoid layer, and also to preserve the normal continuity of the subarachnoid space as well as possible. If the tumor is intramedullary, or if part of an extramedullary tumor must be left behind, it is usually wiser to leave the dura open as a decompressive measure. No harm will come from doing this and often it will serve as an extremely valuable procedure to avoid excess swelling of the cord. Gel film may be used to cover the dural opening in these cases.

Postoperative Care

In any subject such as spinal cord tumors in children, it is impossible to be as complete as one would like in discussion of all phases of the problem. Nevertheless, it seems advisable to say a word or two about postoperative care in the management of spinal cord tumors. The important factors here are to avoid the usual complications which one may encounter. These include, in order of frequency, respiratory complications, skin complications, bladder complications, and phlebitis.

Fluids must be maintained at an optimal level in keeping with the patient's weight and are discussed fully in the chapter on pre- and postoperative care. It is customary to determine frequent blood chemistries on these patients, approximately every other day, so that any abnormal change in nitrogen balance may be immediately detected and accounted for. Patients are routinely turned from side to back to side every two hours, day and night. It is important to aspirate these little patients frequently, and make sure there is no vomitus present in the trachea. Obviously, one must be continuously on the lookout for a possible atelectasis or early pneumonia. As early as possible, these patients are placed on oral diets. The advantage of this is that it is much easier to administer satisfactory amounts of protein via the oral route than any other way. Patients are placed on parenteral vitamins, if necessary, and, if not, on some multivitamin preparation giving them about three times the minimal normal daily requirements The skin is kept scrupulously dry and constant attention is paid to the heels, trochanters, and sacral areas which are most frequently involved from pressure. If there is a paraparesis or paraplegia present, it is extremely important that one institute physiotherapy at the earliest possible moment. Ambulation and constant practice by the patient are the keynotes to re-educate the limbs.

For the same reason that these tumors are difficult to diagnose in their early stages, namely, the changing neurologic structure of these patients as they develop, it is also easier for them to recover from a paraparesis or a paraplegia than it is for an adult.

The problem of proper bladder handling has already been discussed with regard to early insertion of the Foley catheter and placing these

patients on tidal drainage, if necessary. Frequent and serial cystometrograms (once every 10 to 14 days) should be done to evaluate bladder improvement where there is an atonic or cord bladder, both before and after the patient comes to surgery.

Final Results

Figure 11 shows the ultimate outcome in the Pennsylvania series of patients. Only two patients suffered an operative mortality. One of these was an intramedullary tumor in the high cervical region, and the other was an epidural spinal abscess of massive nature, involving the entire cord

FINAL RESULTS OF SPINAL CORD TUMORS IN CHILDREN

Number Malignant	12
Number Benign	18
Operative Mortality (30 Days)	2
Case Mortality	11
Working or Asymptomatic at End of 5 Years	9

FIGURE 11. The surgical outcome in thirty spinal cord tumors of children.

up to the cervical area. Both patients were critically ill preoperatively, and perished of respiratory failure.

A five year follow-up revealed that nine patients were totally asymptomatic. This includes full time school attendance in some cases. Considering the fact that eleven patients were paraplegic before surgery, the prognosis in these tumors seems far from hopeless. Certainly the eventual outcome will depend, to a great extent, on an early and rapid diagnosis. *However, a certain factor of chance plays a part, for regardless of the early diagnosis,* it will be necessary for the patient to have a favorable type of tumor from the pathologic point of view.

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CHAPTER XIV

Tumors—Peripheral and Sympathetic Nerves

I. Joshua Speicel

TUMORS OF THE PERIPHERAL NERVES

NEOPLASMS of the peripheral nerves are extremely rare in children but clinically, surgically, and pathologically they do not vary from those in the adult. They may develop on any nerve—be single or multiple, benign or malignant. The clinical features vary only in regard to the nerve afflicted. Peripheral nerve tumors arise from perineurium, endoneurium, or Schwann sheaths, but not from axis cylinders. Tumors originating from Schwann sheaths are known as neurilemmomas, those from perineurium or endoneurium as neurofibromas. The malignant counterparts of the latter are known as neurofibrosarcomas. Tumors of peripheral nerves, as well as such other neoplasms as lipomas, gliomas, angiomas, pigmented moles and chromaffin tumors, may all be part of the neurofibromatosis complex.

Neurilemmoma (Neurinoma, Schwannoma)

Neurilemmoma is a variety of tumor believed to originate from Schwann sheaths. Since Schwann sheaths invest nerves just as they leave the spinal cord or brain their absence precludes the development of neurilemmomas within the central nervous system proper. Neurilemmomas may occur at any age and are more common in females than males, more often single than multiple, and are most often not associated with other stigmata of Von Recklinghausen's disease. However, the finding of one such tumor should always stimulate a thorough search for others. They are uncommon in children and we have encountered none in the group of 0-14 years of age.

Neurilemmomas may originate either from an intracranial, intraspinal or peripheral nerve. Of the intracranial nerves, the auditory nerve is, with few exceptions, the only one likely to be involved. We have seen such a tumor of the glosso-pharyngeal nerve. Apparently arising from the Schwann sheath cells before the nerve leaves the cranial cavity, the tumor grows

within the posterior cranial fossa where it tends to compress and distort the area of the cerebellopontine angle. A detailed review of these lesions is given in the chapter on brain tumors.

Peripheral neurilemmomas, in a review by Stout, originate most commonly from the large nerves of the flexor aspects of the limbs, as well as



FIGURE 1. Neurilemmomas. Note prominent palisading.

occasionally from the peripheral nerves of the hands, face, scalp, tongue, stomach and chest. Unless the lesion is partially in the spinal canal, or on an intracranial nerve trunk, it will rarely reach the attention of the neurosurgeon. Frequently these peripheral nerve tumors present themselves as intrathoracic tumors as reported by Godwin *et al.* These tumors grossly are well circumscribed and intimately associated with their nerves of origin. However, as a rule, they are well encapsulated so that the involved nerve can usually be dissected free of the tumor. If secondary changes such as edema and cystic degeneration have not occurred, the tumors are moderately firm and grey to pink-grey in color.

There are two most commonly recognized and distinctive microscopic pictures. The Antoni type A is the most characteristic. In this variety, the tumor cells are usually arranged in prominent and striking palisade formations as shown in Figure 1. At other times, atypical palisades must be searched out. The tissue between the palisaded nuclei is eosinophilic or polychromatophilic and rich in collagen. The nuclei are regular and have round blunted edges. The Antoni type B variety on the other hand, shows a disorderly array of tumor cells which vary in size. Collagen fibers in the type B variety are scant in contrast to type A.

Neurilemmomas are invariably benign. They may recur locally if incompletely excised but do not metastasize. Clearly therefore, a determined effort to remove them completely must be made if surgical operation is undertaken.

Neurofibromas and Neurofibromatosis

Neurofibromas may be single or multiple and originate anywhere along the course of a peripheral nerve after it has acquired peri- and endoneurium. There is still considerable disagreement about the origin of these tumors—some claiming a Schwannian origin, others a perineurial origin, and still others an endoneurial origin. It is our belief that they arise from endoneurium or perineurium.

Tumors most proximal to nerve root origins, immediately at the neuromedullary junction, may be partially within the medullary canal and protrude through the intervertebral foramina to form so-called dumbbell tumors according to Griffin, *et al.* and Svien, *et al.* Subcutaneous or dermal nodules as well as growths along any of the large nerves are common. A detailed description of the clinical picture of intraspinal neurofibromata is present in the chapter on spinal cord tumors.

Grossly the neurofibromas are poorly encapsulated and appear to infiltrate the involved nerve in contrast to the previously described neurilemmonas which are well encapsulated. The involved nerve frequently appears swollen

and distorted in a bizarre fashion. It may also appear to be partially replaced with an infiltrating spindle-shaped tumor. Occasionally, when the tumor arises from a smaller nerve branch, the tumor may appear pedunculated. On section, neurofibromas are grey-brown and rubbery to firm. Larger tumors, particularly those extending into the medullary canal may outgrow their blood supply and show cystic degeneration and other secondary changes such as hemorrhage. Microscopically the tumors present a variable picture. The most common, however, is a disorderly array of spindle-shaped cells with deeply stained round to oval nuclei. The nuclei characteristically show considerable variation in size, as well as staining quality. Despite this nuclear pleomorphism, the majority of these tumors are only locally invasive and although they may recur after incomplete removal, they will not metastasize. The gross picture of a distorted, grotesque nerve bundle is frequently also seen under the microscope. The irregularly arranged nuclei are found growing in a fibrous or loose fibromucoid stroma, and the individual cell borders are frequently indistinct. Mitoses are rare. The poor demarcation of the tumor from surrounding tissue frequently makes complete excision difficult or impossible without sacrificing the involved nerve or other tissues in which it is found. In such instances, the signs and symptoms are those of a space occupying, expanding, intrathoracic lesion without neurologic involvement.

If, as stated above, the lesion is partially in the spinal canal, the patient will begin to develop evidence of an intraspinal space occupying lesion with involvement of the long spinal tracts. Not infrequently, before sensory level and motor involvement become apparent, the patient may complain of pain, paresthesias and possibly numbness radiating along the course of the involved nerve root. Since these lesions are generally very slowly growing, radiating nerve root pain may be present for a considerable period of time without any signs of neurological involvement. Such patients may be thoroughly investigated by every neurologic means, including myelography without the source of their discomfort becoming manifest. Such unfortunate patients are frequently called psychoneurotic and dismissed by the neurosurgeon as impossible to treat. We have had occasion to remove such lesions surgically from three different patients, all of whom had been previously discharged as being psychoneurotic. In two of the cases, planographic x-ray studies of the intervertebral foramina showed enlargement of the foramen at the suspected site of the lesion. All three patients were cured following surgical removal of the tumor.

Malignant Tumors

Malignant tumors are said to arise, not infrequently from neurofibromas, particularly after unsuccessful attempts at complete removal. Whether the

malignant tumor may arise from one which has been previously benign in an involved subject, thoroughly discussed in pathological treatises. It is our opinion that fibrosarcomas, neurofibrosarcomas or neurogenic sarcomas were probably malignant to begin with and did not arise from a previously benign tumor. The infiltrative nature of the neurofibroma (as also seen in the dermal nevus), the nuclear pleomorphism, and the local recurrence after incomplete removal are suggestive characteristics of malignancy. Subtle histologic differences between the benign and malignant neurofibroma may exist but be difficult, if not impossible, to recognize.

Fibrosarcomas, Neurofibrosarcomas and Neurogenic Sarcomas show an exaggerated pleomorphic picture of their benign counterpart, neurofibroma. In addition to a greater degree of invasiveness as seen grossly, they may metastasize. The microscopic picture is also variable, depending upon the predominance of fibrous, fibro-myxomatous or neurogenic elements. Grossly the tumors are firm, to soft and cystic, grey to pink or frankly hemorrhagic. The tumors may be very cellular, with numerous spindle-shaped pleomorphic cells in a scant stroma. These may show prominent palisading of nuclei. There are variable numbers of mitoses and nuclear pleomorphism commensurate with the degree of malignancy. Other tumors, softer and more gelatinous, may show areas of myxomatous-like material with stellate myxoma cells as well as fibroblasts. Special stains will demonstrate variable amounts of collagen as well as argentophilic fibers. Except in instances of proven Von Recklinghausen's disease, it is our opinion that any and all neurofibromata should be surgically attacked and removed completely, if at all feasible. Immediate frozen section should be made at the time of operation and if the suspicion of malignancy exists the principle of wide excision of surrounding structures must be followed. If the section unequivocally shows malignancy with mitotic figures, amputation of the involved extremity and resection of all draining lymphatics and lymph glands becomes a fundamental necessity. The only contraindication to the latter procedure is the presence of known or proven, distant and multiple metastases. We have encountered several such malignant lesions but none in the pediatric age group. The youngest seen was in a seventeen year old male with neurofibrosarcoma of the sciatic nerve. Curiously enough, the clinical picture in this young man was that of severe radiating pain down the posterolateral aspect of the thigh, leg and ankle. There was a complete absence of neurologic findings except for the fact that his ankle jerk was entirely absent. By the time he had reached our attention, he had already had a myelogram because rupture of the intervertebral disc was suspected. Actually, the tumor was easily the size of a man's fist and occupied the entire sciatic nerve just below the gluteal fold so that the mass was clearly evident to visual inspection. Although amputation and wide dissection of surrounding

tissues was made this patient died of multiple metastases 6 weeks after he was first seen for these complaints.

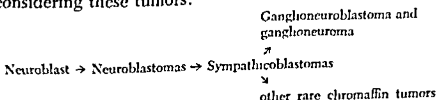
Traumatic Neuromas

Traumatic neuromas are not truly tumors. They appear to be disorganized overgrowths of axis cylinders, neurilemmal sheaths and perineural sheaths intermingled with true scar tissue. They form on the proximal end of a severed nerve trunk or along the course of an intact but injured nerve trunk. If regeneration of Schwann cells and axis cylinders results in the microscopic picture of tangled spaghetti-like masses of nerves and collagen, the names plexiform neuroma or proximal neuroma are frequently used. The distal end of a completely severed nerve develops a smaller mass, composed of gliomatous tissue and is known as the distal glioma. Resection of both the "proximal neuroma" to viable nerve fibers and the "distal glioma" to patent axis cylnders *is mandatory before suturing of a divided nerve is* undertaken. The clinical features, diagnosis, and surgical treatment is discussed under the chapter of peripheral nerve injuries.

TUMORS OF SYMPATHETIC NERVOUS SYSTEM

Tumors originating from sympathetic nervous tissue are common in those of the pediatric age group as compared to the frequency found in the adult, but nevertheless are a relatively infrequent tumor. Almost without exception, such clinical problems are treated surgically by the general surgeon and the neurogenic origin of the tumor is determined after the pathological study has been completed.

Sympathicoblastoma and Neuroblastomas are closely related tumors, probably originating from the same stem cell—the primitive neuroblast or primitive sympathetic tissue. If the cells are slightly more mature and form rosettes and neurofibrils, the name "sympathicoblastoma" is applied. Surprisingly enough, there are cases on record where these highly anaplastic and primitive tumors have gone on to maturity with the formation of numerous small ganglioneuromas or even neurofibromas despite the presence of distant metastases. We have on record several instances of a neuroblastoma discovered in infancy or early childhood with proven metastases confirmed by biopsy, which have gone on to maturation after x-ray therapy. These patients, curiously enough, have survived as reported by Uhlman and VonEssen. From practical purposes, the following scheme may be used in considering these tumors:



Neuroblastomas and Sympathicoblastomas

These tumors are by far the most common malignant tumors in infancy and childhood, and may even be found in the newborn. Approximately 75% of the cases of neuroblastomas are found in children under the age of four. Neuroblastomas may be of such size when they develop before birth that

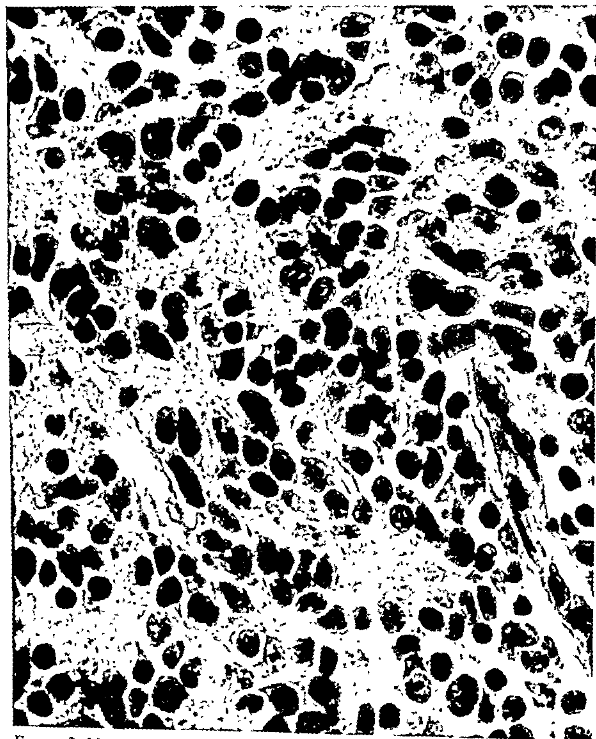


FIGURE 2 Metastatic sympathicoblastoma from the suprarenal with many prominent rosettes.

dystocia results. These neoplasms are sometimes discovered only after metastases have occurred. If the child presents symptoms referrable to bone metastases, particularly to the calvarium, the tumor may be classified as the "Hutchinson" variety: when the first metastases are to the liver, the "Pepper" variety. Tumors occurring in older children are more commonly of the Hutchinson variety (ages 2-8). This entire group of tumors (neuroblastomas, sympathicoblastomas and gangliomas), originating as they do from sympathetic or neural cells, may originate in the suprarenal medullas, or any of the sympathetic nerve plexuses. Grossly, they resemble hematomas. Microscopically, the neuroblastoma consists of small round or spindle-shaped cells with deeply stained nuclei and scant cytoplasm. The cells may easily be mistaken for lymphocytes. In the neuroblastoma, a much more rapidly growing tumor, rosettes and nerve fibrils cannot be demonstrated but as in Figure 2, are common in sympathicoblastomas, slower growing neoplasms whose individual tumor cells have more vesicular nuclei and scant cytoplasm. Combinations of the two varieties, sympathicoblastoma and neuroblastoma, may be found in the same individual.

In the Hutchinson variety of neuroblastoma, the skeletal metastases may stimulate periosteal bone proliferation. The x-ray, as well as subsequent histologic examination, present pictures easily confused with the Ewing tumor. Many so-called Ewing tumors, so diagnosed from bone biopsies, have been shown by follow-up studies and autopsies to be metastatic sympathicoblastomas.

Ganglioneuroma

Ganglioneuromas may be found in the adrenal medullas or in the paravertebral plexuses, nerve roots and splanchnic and visceral plexuses. They consist of ganglion cells, frequently containing intracytoplasmic pigment and nerve fibers. Schwann sheaths may or may not be present.

Grossly, these tumors may be well or poorly encapsulated. On section, they are grey or tan, and may show areas of secondary change, particularly in regard to growth. Cysts, edema and occasional focal calcification can be present.

The malignant variety *Ganglioneuroblastoma* is intermediate in degree of malignancy between the sympathicoblastoma and the more mature ganglioneuroma. Although metastases may occur, the tumor may remain localized. It presents spindle-shaped, more mature appearing cells, some suggesting Schwann cells, endoneurium, sympathicoblasts and multinucleated cells reminiscent of ganglion cells.

Miscellaneous Tumors

Glomangiomas are extremely rare in children and are mentioned only in passing. They are usually benign tumors of the neuromyoarterial glomus.

Such structures may be found in the fingernail beds, about the joints, and the coccyx (coccygeal body tumor). They are extremely vascular and show peri-vascular aggregates of small cuboidal or polyhedral cells with clear cytoplasm and centrally located nuclei. We have not encountered such a lesion.

Chemotectomas (Nonchromaffin Paragangliomas) are tumors arising from structures such as carotid bodies, glomus jugulare, or tympanicus. Tumors such as these are also very vascular. These tumors may be present as hemorrhagic masses in the external auditory canal or in the middle ear. Microscopically the tumor cells closely resemble nerve cells or cells of the adrenal medulla, usually without any pigment. Histologically, these tumors are benign, but because of their location, in the middle ear or temporal bone, complete excision is difficult if not impossible. Almost invariably this problem presents itself to the otorhinolaryngologist. Those cases recorded in the neurosurgical literature have been adults.

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CHAPTER XV

Trauma—Intracranial and Cranial

PHILIP D. GORDY

HEAD INJURIES in children constitute a very important and constantly recurring problem to the pediatrician, general physician, and neurosurgeon. Head injuries in children present themselves with increasing frequency as a result of the mounting traffic and automobile accident problem. An alarming clinical picture may develop with great rapidity and is, of course, a great concern to the parent and physician alike. The potentialities of the injury range from the immediate implications of the injury and its emergency treatment to the problem of future outlook and long range evaluation. Since these injuries may be treated, at least initially, by physicians in many fields, it is of the greatest importance that the problem be clearly understood in all of its aspects.

The child who has suffered a head injury differs in several important respects from an adult who has been subjected to head trauma. The developing child is a much more labile organism. What appears to be a relatively minor blow to the head may produce the picture of a child who is pale and unresponsive. Seizures may develop and the child frequently vomits. The vital signs may be extremely labile. These signs require the utmost in vigilance in order to determine whether they indicate a developing clot with a need for urgent surgery or cerebral contusion and edema. This clinical picture may change rapidly to one of marked improvement with increased responsiveness and more stable vital signs.

The reverse is often seen. It is a common experience to see a child who has fallen from a height, such as an upstairs window, and who appears to have no significant degree of injury. In either event the clinical picture can change with great rapidity. Once improvement has begun the child may improve more rapidly and often with less residual deficit than the adult.

A developing child is, of course, first exposed to the possibility of injury to the head during the process of birth. Excessive moulding of the skull may occur and either mild or severe hemorrhage and fatal termination. It has been shown that blood may appear in the spinal fluid of the newborn as a result of small hemorrhages occurring either into the ventricular cavity or into the surface and basal subarachnoid pathways. Petechial hemorrhages

throughout the white matter have also been shown to occur as a result of the birth process. Clinically, the newborn infant, with a significant cerebral injury may be rather pale and listless with poor respirations. Convulsions frequently are seen. Diagnostic spinal tap will usually reveal blood in the subarachnoid space. Bleeding into the subdural space may be revealed if subdural taps are carried out. Subdural collections incident to birth trauma may, of course, not give signs of their presence initially and may only be detected later when the child fails to feed properly, is irritable or restless, and shows a bulging fontanelle. Subdural tap at this time may disclose a typical subacute or chronic collection of xanthochromic fluid or old blood. The special problem of subdural hematomas in infants is discussed fully in a subsequent chapter.

Recent studies by Foltz *et al.* in 1956 have pointed to the probability that subarachnoid bleeding may so interfere with circulation and absorption of cerebrospinal fluid that communicating hydrocephalus may result, following a severe head injury in an infant or young child. One should, therefore, be alert to detect the early signs of a developing hydrocephalus.

The possibility of a more long range sequela of cerebral birth injury resulting from excessive moulding of the head is suggested by Penfield. Incisural sclerosis is thought to result from squeezing of the medial edge of the temporal lobe over the tentorial edge during the passage through the birth canal. This results in ischemia and resultant gliotic changes in the mesial temporal lobe with the possibility for the later development of temporal lobe epilepsy.

Initial Observation

Closed head injuries may, as in adults, be of either mild or severe degree. One of the striking aspects of injury to the head in children is the rapidity with which an alarming clinical picture may occur. After a blow to the head, the child may be pale, listless, poorly responsive, restless, and frequently will vomit. These signs may be present whether the injury is of mild or severe degree. For these reasons, the child must be observed in a most careful and attentive manner. The physician who first examines the patient with sufficient care and attention to detail, may note the presence of pupillary abnormalities such as rapid variations in the size of the pupils and reactivity to light. Pathologic reflexes such as the Babinski response may be transiently present. The patient's state of responsiveness may change with great rapidity.

Few hospitals are so arranged, that all of the many mild head injuries could be admitted for observation, nor is this necessarily desirable. The child should be carefully checked by a physician and where the symptoms have been mild and have totally cleared, the parents can be instructed to observe the child carefully. The child should be awakened periodically to

make certain that stupor and coma have not developed. Untoward signs, such as vomiting, pupillary change or weakness of one side should be watched for and called to the attention of the physician at once should they occur. If, on the other hand, the child makes an uneventful and progressive recovery the vigilance can be relaxed and activity can be gradually increased. There is no indication for weeks of bed rest and restricted activity as was once advised.

Only the mildest of closed injuries can be handled in this fashion. It is far safer to admit to the hospital the child who has had any significant degree of injury to the head for at least a short period of observation. In these children, therefore, as in the more severe closed head injuries, the child will be admitted to the hospital for continuous, close observation and active treatment measures. The initial diagnostic and treatment measures are carried out in the emergency room or admitting department. It is of the utmost importance that initial observations and an accurate history of the injury be obtained and recorded. Whether or not the child fell down stairs, was struck by an automobile, or was riding as a passenger in an automobile involved in an accident will be of importance in assessing the type and severity of injury which may have occurred. Later medicolegal importance may depend on the accurate recording of the initial history and examination findings.

A rapid, but careful and detailed, physical examination must be promptly carried out. The presence or absence of associated fractures must be noted and proper positioning and splinting performed. Palpation, percussion, and auscultation of the chest are important in determining the presence or absence of rib fractures, and a pneumo or hemo thorax. Abdominal examination should be carried out promptly in order to detect the possible early signs of a ruptured hollow viscus or a rupture of the spleen. The child should be carefully observed for signs suggesting the presence of developing hemorrhagic shock such as might occur from a rupture of the spleen, retro-peritoneal hemorrhage, or hemorrhage associated with major long bone fractures.

The presence or absence of lacerations of the extremities, trunk, or head should be noted and if there is any significant degree of hemorrhage, this should be controlled by appropriate emergency measures and a sterile dressing applied until more definitive treatment can be carried out. It must be remembered that a closed head injury of itself rarely causes a picture of hemorrhagic shock. It is therefore of the utmost importance that a careful search be made for the presence of associated injuries.

The priority of treatment of the case presenting multiple injuries is a matter of careful clinical judgment. Too often, major associated injuries may be delayed in treatment because of a time honored tradition that even a

mild head injury must take precedence. Many times this will be so. However, even in the presence of a major closed head injury, the obvious signs of developing intra-abdominal hemorrhage or obvious peripheral respiratory distress will demand immediate measures to control these situations. The neurosurgeon who detected the signs of a developing epidural hemorrhage would not hesitate to proceed immediately with major surgery to prevent a fatal outcome. In the same way, major associated injuries which might result in a rapidly fatal outcome must be handled with priority.

In general, less serious associated injuries, may be deferred for a time until the patient's condition has stabilized and it is obvious that no major intracranial problem is developing. However, one should not defer lacerations requiring debridement, cleansing, and closure, compound fractures, or fractures with displacement for too long a period of time. Otherwise, permanent and lasting damage may occur when the child actually would have withstood judicious intervention at an earlier time.

Initial accident room care will also in many instances require prompt and active measures to secure an adequate air-way and sufficient oxygenation. The child should not be allowed to remain on its back with mucus and secretions as well as vomitus draining back into the pharynx and obstructing the trachea. If not contraindicated by associated injuries, the child should be turned on his side in order that secretions and vomitus may drain out of the mouth by gravity. Oxygen by nasal catheter should be at hand for administration once the air passages are clear of any obstructing secretions. One should not hesitate to perform immediate tracheotomy if it is not possible to clear the air-way adequately by intratracheal suction and to maintain it in an obstructed fashion as advocated by Dunsmore, *et al.* The tracheotome devised by Sheldon *et al.* may be used in the early emergency care of severe head injuries.

If shock is present, prompt measures for correction should be carried out. An intravenous infusion should be started immediately with glucose in water. Neutralized emergency blood may be used initially but typing and cross match should be carried out promptly if there is a need for further blood replacement. Oxygen, of course, should be administered. Morphine or demerol should not be used because of their possible depressant action and pupillary effects.

Concurrently with the assessment of the patient's general situation, and attention to the most urgent emergency measures for survival, certain important neurologic observations will have been made. A complete and detailed neurologic examination will ordinarily not be possible. However, the restless, stuporous child should be observed to determine whether or not he moves both sides and, if he does so, whether or not they move equally. The tone of the extremities is important. If there are no extremity injuries, the

arms and legs should be moved and the presence or absence of flaccidity or spasticity determined. The child may present the picture of a decorticate or decerebrate rigidity. The patient may develop deep and stertorous respirations with rigid extension of the lower extremities and either *flexion or* extension whenever stimulated. The presence of this neurologic picture indicates a severe degree of injury to the central nervous system and may point to either a decompensating intracranial hemorrhage, a severe brain stem contusion or bilateral, widespread, cerebral contusion. The evaluation of motor activity, in the comatose or semi-comatose patient, will provide the most important information regarding the general state of responsiveness. Observation of the patient when he is requested to move the extremities will indicate whether or not there is a sufficient level of consciousness to carry out this command. The patient may on the other hand respond only to painful stimulation and may show no spontaneous movements. Supraorbital, Achilles tendon pressure or deep muscle pressure may be used to elicit movements. Only after such stimulation may one be aware that there is a paresis of one side.

Careful observation of eye signs is another important source of diagnostic information. In the presence of severe brain stem injury the pupils may be either dilated and fixed to light or constricted. The presence of such eye findings may indicate a major brain stem injury with a poor prognosis. Occasionally, the pupils may be seen to rapidly dilate and constrict as the condition of the patient alters. The presence of pupils which are equal and react to light suggests a better prognosis in the absence of signs to the contrary. A unilateral, widely dilated and fixed pupil usually indicates an urgent, surgical situation and results from tentorial herniation of the uncus with pressure on the adjoining third nerve. There are, of course, instances in which such a third nerve palsy results only from direct contusion or stretching of the nerve and does not indicate a developing hemorrhage. One should not assume this to be the case, however, until a rapidly developing hemorrhage has been ruled out by appropriate surgical intervention.

If the dilated pupil is observed when the patient is first seen a half to several hours after the accident, regardless of whether the patient is conscious or not, the patient probably has a basilar skull fracture with direct contusion to the oculomotor nerve and can be further observed withholding operation.

Forced conjugate deviation of the eyes may indicate either a destructive lesion in the region of the frontal eye fields with paralysis of conjugate gaze away from the side of the lesion or an irritative lesion with forced conjugate gaze toward the opposite side.

Funduscopic examination is important in establishing a baseline for later

observation. Papilledema, of course, will not be seen in the acute lesion. However, a normal fundus on admission, which changes over the course of several days to the picture of choking of the optic disks, is of great significance and may indicate the presence of an expanding intracranial hematoma. In addition, a severe, closed head injury may produce sufficient subarachnoid bleeding to result in hemorrhages within the retina. These hemorrhages have important diagnostic significance and indicate the presence of subarachnoid bleeding and, therefore, cerebral contusion and laceration. A lumbar puncture performed in a patient with such a retinal finding will invariably reveal bloody spinal fluid.

In evaluating a child with a head injury, the patient's general state of responsiveness is probably the most important single indicator. Nurses and House Officers should be thoroughly familiar with the importance of judging the level of responsiveness and of methods of evaluating it. If the child responds to questions or commands to any degree, a level of responsiveness is established from which a later trend can be determined. The response may be slight such as a movement of the eyelids in response to a request to "open your eyes," or the child may groan or attempt to answer in response to the use of his name. If there is little or no response to voice, painful stimulation will enable one to determine the response level. Simple methods of eliciting such a pain response are supraorbital pressure, Achilles tendon pressure, or deep pressure on the pectoralis muscles. In this, as is the case with other observations in the comatose or semi-comatose patient, it is the dynamic trend of the picture, rather than a single observation which is of importance. The level of responsiveness may well prove to be a more sensitive indicator of a deteriorating neurologic status than a change in the graphic picture of the vital signs.

An essential part, of course, of the initial neurologic evaluation is the start of an accurate graphic record of the patient's blood pressure, pulse, temperature, and respirations. Blood pressure, pulse, and respiratory rate observations should be kept at frequent intervals on a graphic record so that one can easily see at a glance whether or not there is any significant trend in the vital signs. The frequency of the observations will depend on the individual case and will, of course, need to be altered depending on the condition of the patient. Rectal temperature observations may be indicated at two hourly intervals. Again, it is the trend which one may see as a result of repeated observations which is of importance rather than the level of any single blood pressure or pulse observation. The classic picture of an acute rise in intracranial pressure characterized by a slowing pulse, rising systolic and falling diastolic pressure to produce a widened pulse pressure, and slowing respirations is, of course, an indication of developing intracranial

pressure with the need for urgent surgical intervention. One single observation, however, such as a slow pulse may need to be evaluated, taking into consideration the remainder of the patient's clinical condition. One may occasionally see a very slow pulse in a patient who is rapidly clearing and whose other signs do not indicate a deteriorating state resulting from rising intracranial pressure. It is wise to observe such a patient carefully, being prepared to intervene surgically if necessary, even though the possibility exists that the slowed pulse may not indicate the presence of a space occupying clot.

It must be pointed out that the so-called classic picture of an acute rise in intracranial pressure may actually be a late phenomenon. It is for this reason that evaluating the total picture is of such tremendous importance. A depression of the level of responsiveness may be an earlier indicator of a beginning deterioration. If one is content to sit by until the so-called "focal signs" develop or the classic signs of increased intracranial pressure make their appearance, there will be occasional cases which are found to be irreversible in spite of surgical intervention at this time.

The deep and superficial reflexes should be carefully checked and reevaluated at intervals. The appearance of a unilateral hyperreflexia when the initial examination revealed the reflexes to be equal is of definite significance and may indicate the clinical onset of an expanding intracranial hematoma. The presence or absence of pathologic reflexes such as the Babinski response and the Hoffman reflex may also be of importance in indicating a change in the neurologic picture.

If one is therefore careful to evaluate the neurologic status of the child in this fashion, a fairly accurate picture of the extent of neurologic damage may be formed and appropriate treatment may then proceed on a rational basis. The treatment of an acute head injury divides itself naturally into two general phases. The first, or observation phase, is directed toward the gathering and recording of careful observations of the patient's neurologic status in order that any signs indicating a need for surgical exploration may be detected promptly. This is one of the most important reasons for the careful recording of the patient's initial status and for the unremitting observations outlined above. The second general aspect of care is the medical management of the contused and swollen brain in the absence of a lesion requiring urgent surgical intervention. This treatment may be conveniently divided into several categories.

Non-operative Floor Care of the Acute Head Injury

Maintenance of an Adequate Air-way

The maintenance of a clear air-way is of paramount importance in the treatment of any head injury. The patient who is in a stuporous or comatose

state should not be permitted to lie on his back, thus permitting the tongue to fall backward and secretions to drain back into the pharynx and trachea. The child should be placed on the side with the foot of the bed slightly elevated. With the head and face turned somewhat toward the bed, secretions and vomitus will naturally drain out of the mouth and will not be aspirated. The nurses and house officers should be carefully instructed in the performance of proper intratracheal suction to be carried out at frequent intervals in order to keep the tracheo-bronchial tree free of secretions. Should it be difficult or impossible to achieve adequate intratracheal suction there should be no hesitancy in performing tracheotomy. Tracheotomy should be used before it becomes an emergency. One might make the observation that an opening in the trachea may in many instances prove more life saving than making an opening in the skull!

Oxygen by nasal catheter or mask should be given in the acute head injury in either a comatose or semi-comatose state. A higher percentage of oxygen concentration can be given by either of these methods than by tent. These methods are also preferable to an oxygen-tent, since in the tent the patient either gets oxygen at the expense of nursing care, or nursing care at the expense of oxygen!

One cannot overestimate the importance of a clear air-way with adequate oxygenation of the comatose or semi-comatose patient. Anoxia or hypoxia of any significant degree will certainly prejudice the patient's chances of recovery by establishing a vicious cycle of cerebral edema and increasing venous congestion.

Sedation

Mild sedation may be necessary to combat extreme restlessness. Small doses of sodium phenobarbital intramuscularly, sodium amytal, or paraldehyde also given intramuscularly are suitable. Care should, of course, be taken to avoid any over sedation which might obscure a trend toward a lowered state of consciousness or mask important changes in either direction. Small doses of codeine may be used in the presence of associated painful injuries. More recently tranquilizing drugs such as sparine have proven to be of great value by decreasing extreme restlessness with a maximum of safety. The dosage will need to be adjusted to the age and size of the individual patient

The Use of Spinal Tap

The use of spinal tap in the diagnosis and treatment of acute head injuries is still somewhat controversial. The trend, however, at the present time is certainly in the direction of restriction of its use from a therapeutic point of view rather than the use of repeated spinal taps as was once em-

ployed. A diagnostic spinal tap at a time when the child is not excessively restless may prove of value in revealing the presence of blood and of indicating an increase in intracranial pressure. The spinal puncture should be done carefully with the child as quiet as possible so that an accurate pressure level may be obtained. It is, of course, important to examine the fluid carefully to be sure that any blood contained therein is a result of the head injury itself and not the result of a traumatic tap in a restless patient. It should be remembered that the presence of a normal or only slightly elevated pressure does not necessarily rule out the presence of a space occupying lesion in the cranial compartment. The intracranial pressure may not always be reflected accurately at the lumbar spinal needle. If there is in the patient's clinical state a suggestion of a significant elevation of intracranial pressure, then the importance of the information to be obtained from a diagnostic spinal tap is probably overshadowed by the possibility of producing a tentorial herniation of the uncus and spinal tap should therefore not be employed. Repeated spinal taps as a therapeutic measure are no longer employed in most clinics. In an occasional case the headache and discomfort occasioned by blood in the subarachnoid space may, in the absence of any signs of an elevated intracranial pressure, be sufficient indication for the performance of several repeated taps in order to withdraw some of the irritating blood. Occasionally, the patient may be made more comfortable in this way.

The Use of Hypertonic Solutions

Various hypertonic solutions have been used over a period of many years to combat acute brain swelling. The use of these substances is much more restricted at the present day, but some of them may prove useful in certain instances. Fifty per cent glucose is one of the most commonly used solutions. This may be given usually in 50 cc. amounts intravenously and may be repeated in four to six hours if necessary. Its use should not be continued over several times, however, because of the possibility of a secondary rise in intracranial pressure. It may on occasion be very effective in controlling an acute increase in brain swelling as may sometimes be demonstrated at the operating table with the brain exposed. Concentrated plasma has been used by many and found to be of some benefit. A standard unit of plasma is mixed with one half the usual volume of water and the unit of plasma then administered slowly intravenously. Salt free serum albumin has also been used. Three per cent saline solution in amounts of 50-100 cc. once or twice daily has been found to be of benefit without a secondary rise in intracranial pressure. Moyer has suggested m/2 molar sodium lactate solution given in amounts of 5 to 7 cc. per kilogram of body weight intravenously. This has been found to be of value in decreasing brain swelling, but its use should

not be continued beyond the need for an emergency measure to control brain edema. Recently concentrated solutions of urea have been used to decrease cerebral edema.

The indiscriminate use of hypertonic solutions may be harmful and their use should be judiciously restricted. Excessive dehydration of the patient is also to be condemned. The patient should have an adequate fluid volume intake in order to cover physiologic needs. The volume will vary depending on the age and size of the child, but if the fluid is given intravenously it should be administered slowly. No significant decrease in cerebral edema occurs from the dehydration regimens which were practiced at one time in the care of acute head injuries.

Temperature Regulation

Hyperthermia is a frequent accompaniment of severe brain contusion and may indicate swelling in or direct damage to thermo-regulatory centers in the brain stem and hypothalamus. The temperature may rise rapidly to extremely high levels. It is important that this temperature rise be brought under rapid control. If the temperature should rise above 103° (R) iced alcohol sponges should be used to prevent a further rise and lower the temperature more nearly to normal. A low enema containing 30 grains of aspirin may be of benefit before the temperature rise has progressed too far. If the temperature continues to rise to higher and higher levels, it may be necessary to use ice packs or an ice-water enema. In children these may be shocking procedures and five to seven minutes is the usual time limit that the ice should remain over the child or before the ice-water enema is siphoned off.

The recent use of hypothermia in the reduction of brain metabolism and control of cerebral swelling suggests that this measure may come to play an important role in the management of a child who has suffered a widespread cerebral contusion with severe cerebral edema. The temperature can be reduced simply by the application of plastic ice bags. The child may then be maintained at the lowered temperature (92-95°F.) until the need for this measure is no longer present. The subject of hypothermia is fully discussed in Chapter V on Anesthesia.

Antibiotics

Ordinarily the use of routine antibiotics is unnecessary particularly if it appears that the state of consciousness may rapidly improve toward normal. However, if the initial examinations or follow-up observations have demonstrated the presence of rhinorrhea or otorrhea, then antibiotic therapy using penicillin or one of the numerous wide-spectrum antibiotics, is, of course, definitely indicated.

Control of Convulsive Seizures

Convulsive seizures are frequently seen as an accompaniment of acute head injuries in children. They may result from either cortical contusion and laceration with subarachnoid bleeding or from blood in the subdural space. Generalized or focal seizures may occur. Prompt control of the seizure should be instituted in order to prevent further cortical damage from hypoxia and the possible onset of status epilepticus. Sodium phenobarbital or sodium amytal intramuscularly may be used. Medication should be given orally, of course, as soon as the patient's condition permits. If seizures are repeated at frequent intervals and it is not possible to regain sufficiently rapid control with intramuscular medication, it may then be necessary to resort to intravenous medication. Intravenous sodium amytal given very slowly with careful observation of the respirations may be effective in bringing the seizure state under prompt control so that intramuscular and later oral medication may be instituted. Rectal avertin and open drop ether have been used in intractable seizure states. One of the most effective methods of gaining prompt control of a series of repeated convulsive seizures is the careful administration of (2½% sodium pentothal) intravenously. This, of course, must be under constant observation, to regulate the depth of depression, by either the attending physician, a responsible house officer, or a member of the anesthesia department.

Recently parenteral dilantin has been placed on the market. This may be given intramuscularly or intravenously in doses of 100-150 mgm. depending on the age of the child. It is effective and does not carry the danger of central nervous system depression.

It is important to remember that a focal or Jacksonian fit in a child does not necessarily mean that there is a focal, surgical lesion. Generalized cerebral contusion may show only such a focal manifestation. Todd's postictal paralysis may be seen following such a series of focal seizures. This may, of course, be of value in indicating a local lesion. The presence of convulsive seizures in the early post-injury stage does not necessarily forecast the development of a later post-traumatic seizure state.

X-ray Examination of the Skull

In a closed head injury it is not necessary or even desirable to obtain an immediate skull x-ray at the time when the child may be extremely restless and the condition somewhat precarious. A trip to the x-ray department can be safely deferred until the child's condition has stabilized, associated injuries have received emergency treatment and shock, if present, has been corrected. It is a distressing experience to see a child with an acute head injury being forcibly restrained on the x-ray table, crying and thrashing while an attempt is made to obtain x-rays which are almost inevitably not of diagnostic quality. Due to an unfortunate public misconception of the

immediate necessity for skull x-rays a few words of explanation may be necessary in order to inform the parents of the reason for deferring the x-ray examination of the skull. In cases involving depression of the skull or penetration into the calvarium, skull x-rays are, of course, extremely important and must be obtained as soon as the condition of the patient permits.

Fluid and Electrolyte Balance

Initially, the patient's nutritional requirements are in most instances satisfied by the intravenous administration of an appropriate volume of 5 or 10% glucose in water. The solution should be administered slowly and of a volume appropriate to the normal fluid requirement of a child of that age. No value, and indeed harm, may come from attempts at dehydration by restriction of fluids. If there is not a prompt restoration of consciousness so that oral intake can be substituted for the intravenous route, then a gastric tube should be inserted for the administration of a proper fluid volume and caloric requirement. Several commercially prepared formulas are available. However, one of the most satisfactory formulas is the following which can be prepared in any hospital diet kitchen:

1 or 2 qts. whole milk
2 or 1 qts. tap H_2O
2 tablespoons sugar
2 teaspoons salt
1 teaspoon sodium bicarbonate
3 0 Gm. KCl.
60 drops Visyneral
2 eggs

This formula is somewhat low in caloric value, and therefore should not be used over any lengthy period of time without additional supplementation. It has the advantage of not producing diarrhea which is a troublesome objection to many formulas. The tube should, of course, be changed weekly and withdrawn as soon as the patient is able to take nourishment orally.

Mineral and electrolyte requirements should be carefully watched and supplements can be added to the formula as necessary. Baseline electrolyte study should be carried out in any child in whom there is not a prompt return to a normal or nearly normal conscious state. Supplemental vitamin therapy is also of value and may be added either to the intravenous solution in soluble form or added to the tube feeding formula. This subject of fluids and electrolyte balance is fully discussed earlier in Chapter IV, Principles of Pre- and Postoperative Care.

Nursing Care

It is of the utmost importance that staff or special duty nurses to whom the care of an acute head injury is entrusted should be thoroughly versed in the various aspects of the care of such a patient. As a part of her nursing

routine, the nurse should carefully observe the presence of anisocoria, lack of motion of one side or by deepening of the state of responsiveness. She should be thoroughly trained in the methods of carrying out the foregoing treatment procedures such as proper intratracheal suction, the application of iced alcohol sponging or ice packs and careful management of the fluids ordered. The patients should be turned frequently and positioned carefully so that secretions will drain from the mouth rather than be aspirated. Skin care should be meticulous and the use of doughnuts and rings avoided since these may increase rather than decrease the hazard of skin ulceration. Care and time spent in the proper training of responsible nursing personnel will be repaid many times.

Intracranial Hemorrhage

Intracranial hemorrhage is the urgent indication for surgical intervention which may develop as a result of a closed head injury. There are several varieties of intracranial hemorrhage depending on their relationship to the covering structures of the brain.

Extra-dural Hemorrhage

Extra-dural hemorrhage is one of the most urgent neurosurgical emergencies. This lesion may occur in childhood as well as in adult life. The usual picture is that of a child who either falls, striking his head, or is struck by an object such as a baseball or a baseball bat. The child may be rendered unconscious momentarily and then exhibit a return to normal or nearly normal consciousness. He may complain of persisting headache and vomit. From this state he may lapse into a condition of progressively deepening stupor, coma, and, if the lesion is not evacuated, death.

The neurologic picture most commonly seen is that of a child showing progressively deepening stupor and exhibiting a hemiparesis. A dilated and fixed pupil on the ipsilateral side usually accompanies this. A classic alteration in the vital signs may occur consisting of a slowing pulse, a widening pulse pressure, and an elevation in the systolic pressure. It is of importance to note here that the slowing pulse and increasing systolic blood pressure together with the dilated pupil are late, rather than early signs. A dilated pupil is indicative of tentorial herniation with third nerve compression. The slowed pulse, slowing respirations and rising systolic pressure indicate beginning failure of the medullary centers and may in some instances indicate decompensation which is irreversible. A far more sensitive indicator is the deteriorating state of responsiveness exhibited by the patient. On the other hand, the clinical picture may present itself with progressive restlessness, rapid pulse and respiratory rate, and an increasing blood pressure—all indicating a very rapidly deteriorating situation requiring immediate surgical intervention.

Extra-dural hemorrhage most commonly results from a tear of the middle meningeal artery secondary to a fracture line crossing the course of this vessel. This hemorrhage is, of course, arterial and is rapidly progressive. Ordinarily this lesion will have produced profound coma and death if not evacuated within 8 to 12 hours. Occasionally, the time interval may be longer.

The following case history is illustrative of a typical extra-dural hematoma:

Del. Hosp. 252738

N. H., *a little seven-year-old Negro male, was admitted to the hospital on 6/18/57. He was injured when he ran in front of a horseshoe player. The thrown shoe struck him in the left fronto-temporal region. He was knocked to the ground but was not unconscious. He sustained a laceration in the left fronto-temporal region. He was seen in the hospital emergency room where examination revealed a responsive and alert seven-year-old boy.* Blood pressure 104/60-P-104. Respiration normal. A 1½ cm. laceration was present over the left temporal area. After cleansing of the wound a depressed fracture could be felt in this region. The patient moved both extremities equally well and showed no motor or reflex abnormality. The pupils were equal and reacted to light and the ocular movements were normal. There was no papilledema and no hemorrhages were noted. The patient responded promptly and coherently to commands and questions.

Because of the compound depressed fracture of the frontal temporal region, *he was scheduled for surgery approximately one hour later. In the meantime he was placed on careful observations of his state of responsiveness and vital signs. Shortly before the scheduled time of the surgical procedure he suddenly became unresponsive with a dilated and fixed left* pupil. No change was noted in his vital signs. He was immediately placed on the operating table and preparations made for cranial surgery.

After careful preparation of the scalp and debridement of the edges of the wound, the incision was extended in the form of a flap and the depressed area of bone elevated and removed. A large acute epidural hemorrhage which was actively bleeding from a laceration of the middle meningeal artery was found beneath the depressed area. After evacuation of the clot, it was possible to visualize the tear in the middle meningeal artery which was easily controlled with the Bovie. After careful attention to hemistasis in the epidural space, a routine two-layer closure was carried out.

By the completion of the procedure, the patient was beginning to stir about and talk and the pupil had returned to normal size. Thereafter this patient made an uneventful recovery and was discharged asymptomatic with no neurologic findings on his eighth postoperative day.

One must keep in mind that fracture lines crossing major venous channels may also produce serious extra-dural bleeding. A fracture line across the transverse sinus may, for example, produce extra-dural bleeding above the

sinus, overlying the occipital poles, or may bleed into the extra-dural space overlying the cerebellum. This latter site of bleeding produces a recognizable clinical picture permitting evacuation of the clot before an irreversible stage has been reached.

The patient may sustain a fall on the occiput following which a transient loss of consciousness is usually noted. Again there may be a lucid interval followed once more by progressively deepening stupor. In addition, incoordination, slurred speech, nystagmus, headache, and vomiting may be noted. In other words, the patient develops the picture of an acute posterior fossa lesion. Schneider *et al.* (1951) have called attention to the possibility of an extra-dural clot in this location. There are other instances in which cerebellar signs are not present and the patient presents only the picture of deepening stupor. Here, the presence of a scalp bruise or laceration and a fracture line crossing the transverse sinus may be the important clue leading

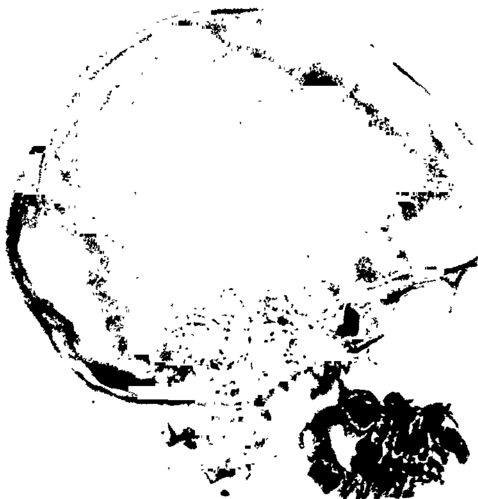


FIGURE 1. X-ray showing compound comminuted depressed fracture of left frontal region with underlying extra-dural hematoma.



FIGURE 2. Post-operative skull x-ray revealing craniectomy site and additional trephine opening.

to the correct diagnosis. In such a situation burr holes should be placed over the cerebellum and a search made for either an extra-dural, subdural, or intracerebellar clot. Prompt evacuation may produce a very satisfactory recovery.

As soon as there is the suspicion of an extra-dural clot immediate operation should be carried out. The approach in the usual middle meningeal hemorrhage consists of making a trephine opening in the temporal fossa of the appropriate side and enlarging this with the rongeur to permit removal of the semi-fluid, semi-solid clot. The clot is removed rapidly to permit visualization of the actively bleeding artery. This may be controlled by dessication with the Bovie, ligated with a suture ligature of fine silk, or, if necessary, the foramen spinosum may be plugged with cotton mixed with a small amount of bone wax. Usually the dura will rise into normal relationship beneath the skull promptly once the clot has been removed. It may be necessary to tack the dura to the pericranium with sutures about the margin

of the exposure and to use gel foam or oxycel pledgets to control residual oozing from the extra-dural space. Frequently it is necessary to turn a small tempero-parietal flap for additional exposure if the clot is quite solid and hemispheric in extent. One should also be prepared to place enlarged trephine openings over the cerebellar hemispheres should the clinical picture and x-ray studies indicate the possibility of a posterior fossa extra-dural collection.

Subdural Hematoma

Hemorrhage into the subdural space is limited in this discussion to those lesions seen in older children. The special problem encountered in infants is discussed in a separate chapter.

Subdural hematoma in the older child differs in no essential respect from that seen in adults. Acute, subacute, and chronic varieties are distinguished. In the *acute subdural hemorrhage* the lesion usually results from cerebral contusion and laceration. The patient may exhibit a pattern of momentary unconsciousness followed by a lucid interval. Again there is a recurring loss of consciousness as is seen with an extra-dural hemorrhage. Progression is usually somewhat slower in the acute subdural hemorrhage though this need not necessarily be the case. The patient may, on the other hand, show no lucid interval but continue to show progressive deterioration. Various neurologic abnormalities may be present depending on the region of maximum focal damage. Hemiplegia or hemiparesis, aphasia, and a dilated pupil may be present. Nuchal rigidity secondary to blood in the subarachnoid space and cervical nerve root irritation may be present. A spinal tap will disclose bloody spinal fluid under increased pressure.

It should be emphasized that the actual subdural bleeding present may not be of major proportions. The brain contusion and laceration, to which the subdural collection is secondary, may be of much greater significance. Realization of this fact is of particular importance in relation to intracerebral bleeding which will be discussed below. However, large, acute subdural collections may be encountered as well and the deterioration may be rapid and profound, as with an extra-dural clot.

A more prolonged and gradual picture results from a *subacute subdural hematoma.* The signs are more gradual and progressive with headache, perhaps unilateral weakness and hyperreflexia and slowly developing stupor. Spinal taps may disclose xanthochromic fluid under moderately increased pressure. Superimposed on this picture at any time may be the familiar signs of an acute rise in intracranial pressure. The clot is usually partly fluid and partly solid and there is frequently a tendency for early membrane formation.

A lesion frequently seen in children and one which gives much the same

clinical picture as the subacute subdural hematoma is the *subdural hygroma.* This consists of a thin, usually xanthochromic fluid present in the subdural space which results in cerebral compression just as does the hematoma. The collection is thought to result usually from a tear in the arachnoid membrane permitting the escape of spinal fluid mixed with small amounts of blood, into the subdural space where it cannot be absorbed. The hygroma is frequently bilateral and there may be a communication between the two sides. The subdural hygroma is perhaps more likely to present with the signs of in-

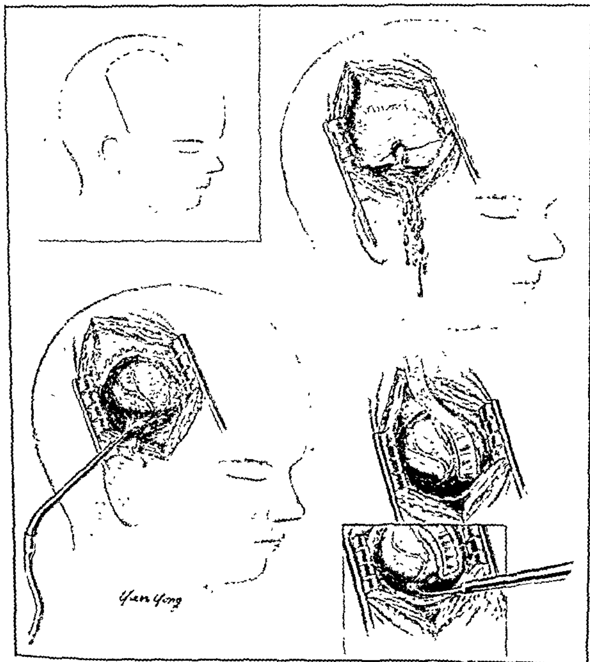


FIGURE 3. Plate showing location of and method of attacking an extra-dural hemorrhage If necessary the oblique incision can be converted into a flap as shown in the insert in order to provide greater room for removal of the clot.

brain tissue. This may produce the picture of temporal lobe herniation or the hemorrhage may, prior to this event, break through into the subdural space producing an acute subdural hematoma. When, on trephine exploration, one encounters a thin layer of subdural clot which does not satisfactorily explain the clinical state of the patient, one must keep this lesion in mind. The subdural collection in this instance is secondary to the rapidly expanding intracerebral clot and prompt evacuation must be carried out.

This case is illustrative of a small depressed fracture of the frontal bone with a more severe underlying lesion consisting of cerebral contusion and laceration with a massive intracerebral hemorrhage, acute subdural bleeding, and intractable convulsive seizures secondary to the above.

St. F. 2643

J. D., Jr. was admitted to the hospital 6/21/57 for evaluation and treatment of a possible subdural hematoma. About twenty-four hours prior to admission this patient was struck on the left side of the forehead in a baseball game. He was unconscious for approximately two hours and then regained consciousness. However, he was constantly drowsy and responded only to painful stimulation or verbal commands in a somewhat incoherent way. He had also vomited frequently. He complained of generalized headache. His past history was non-contributory except that about seventeen days before he had been struck over the same region but at this time was unconscious for only a few moments.

Examination at the time of admission revealed a somewhat lethargic sixteen-year-old boy who seemed to be well oriented and whose recent memory was good. He was able to answer questions well and co-operate. There was an abrasion and swelling over the right side of the forehead. The pupils were equal and reacted to light. The ocular movements were normal No nystagmus was noted. He complained of diplopia on left lateral gaze. The left optic disk was normal and there were no retinal hemorrhages. The right was not visualized. He moved both sides equally well. The grip was equal. The deep-tendon reflexes were equal and physiological and no pathologic reflexes were noted.

The clinical impression was that of a generalized cerebral contusion but he was placed under careful observation for evidence of possible intracranial bleeding. Over the next few hours his condition deteriorated and he became extremely restless and agitated requiring both physical and drug restraint. He became quite incoherent and combative. Examination revealed the pupils to be equal and reacting still. Both sides were moving equally. The pulse was regular and the rate was 68-78. He showed some evidence of dehydration clinically. There were no abnormalities in the vital signs to suggest an expanding clot. Intravenous fluids were started to correct the patient's dehydration and mild sedation was administered.

His condition continued to deteriorate in spite of these measures and it was felt there was probably an underlying space occupying lesion For this reason the patient was taken to the operating room where a right frontal

osteoplastic craniotomy was carried out. A small depression of the frontal bone overlying the frontal pole on the right side was noted. Immediately beneath this the dura appeared to be quite blue and on opening the dura a large amount of degenerated and broken down brain tissue with a small amount of old blood erupted through. A cortical incision was made at this point and a large amount of necrotic brain tissue and partly fluid partly solid old blood evacuated. After all necrotic brain and hemorrhage was removed and all bleeding was under control, the brain was relaxed and under no tension. Following an attempt to close the dura the brain once again became hard and tended to herniate. Wide opening of the dura was necessary together with amputation of the herniating portion. Following this a dural graft was placed using temporalis fascia. The bone flap was then replaced in position and the usual closure carried out. A tracheotomy was performed following completion of the craniotomy.

The patient's immediate post-operative condition was rather critical. However, the extreme restlessness improved and his condition gradually improved until at the time of discharge he was responding well and was asymptomatic. There were no neurologic residuals at the time of discharge. The patient is making a very satisfactory recovery.

Frontal and temporal trephines should be made in the locations previously outlined. If focal signs are present, the exploration should be planned so that the involved side is exposed first. Additional check trephines can then be made after the progressive lesion has been brought under control. An exploring needle is used to explore the subcortical region in the anticipated location. A prompt gush of fluid and clotted blood under pressure plus fragments of softened and necrotic brain tissue discloses the underlying lesion. It may be possible to achieve adequate exposure of the lesion by enlarging the trephine opening with a rongeur. However, it is frequently necessary to use a small osteoplastic flap in order to facilitate the making of a cortical incision and the complete removal of subcortical clot and proper debridement of necrotic brain. Such a subcortical hematoma cannot be properly evacuated by irrigation through the exploring needle. When complete evacuation has been accomplished the brain should be relaxed and under no tension. The most frequent sights of contusion and necrosis are the temporal and frontal poles. A hemorrhage of significant size may lie immediately beneath the surface in the subcortical white matter. Botterell has called attention to the contused and softened brain at the temporal and frontal poles which may require thorough debridement.

Open and Penetrating Wounds

Care of Scalp Lacerations

This aspect of the care of head injuries is all too frequently brushed lightly aside as being of little importance. The most inexperienced house officer is

often illadvisedly entrusted with the repair of a major scalp laceration. Often a laceration requiring extensive cleansing and debridement is closed in the accident room where facilities and assistance necessary to accomplish a proper closure are not available.

Wide and adequate shaving of the scalp must be done. The scalp should be cleansed with soap and water or phisohex. A sterile dressing can then be applied under pressure until preparations for the closure are complete. Adequate debridement of the wound edges must be done in order to remove all devitalized tissue. Naturally, care must be used to avoid sacrificing viable scalp unnecessarily. Bleeding from the scalp edges is easily controlled by galeal hemostats turned back over the scalp edge. Thorough and copious irrigation of the wound with sterile saline solution is then carried out. In the absence of an underlying fracture requiring specific treatment as discussed below, the wound is now ready to be closed. If the galea has not been destroyed, a two-layer closure should be carried out. Fine silk should be used for the galea and the skin may then be closed with interrupted skin sutures of wire or silk.

If there has been extensive avulsion of scalp tissue or if it has been necessary to debride devitalized scalp extensively, it may be necessary, in order to achieve sufficient relaxation of tissue, to perform various plastic maneuvers such as the fashioning of rotation flaps. By extending the ends of the laceration, by undermining the scalp flaps and by the use of appropriate rotation flaps, it may be possible to close a large defect without tension. If the loss has been too great to accomplish this, the rotation flap can be shifted to cover the denuded area and a split thickness skin graft employed to cover the donor site. Difficult technical problems of this nature may arise making desirable the services of a plastic surgeon. These problems are discussed under the chapter on Correlative Plastic Surgery.

Depressed Fractures

Simple Depressed Fractures

Simple depressed fractures will in most instances require elevation. They are, however, not necessarily immediate emergencies. Depressed fractures may be elevated to achieve a better cosmetic result, particularly in the frontal regions, to relieve local or general compression of the brain and to prevent the development of later seizures resulting from cortical irritation. Very slight depressions overlying a silent area may not require elevation.

Pre-operative x-rays are, of course, essential in evaluating the extent of depression and the necessity for surgery. One should bear in mind that there is usually greater fragmentation and depression of the inner table than is evident on the x-ray film. Particular care should be taken in evaluating a depression overlying the longitudinal sinus A slight depression may be

better left alone. Significant degrees of depression will require elevation. However, great care should be taken to insure that all preparations have been made to handle major venous bleeding from the sinus.

In the very young child or infant the so-called "ping pong ball" fracture may usually be elevated by catching the center of the depressed area with a sharp hook and elevating it back into place. More frequently, it is necessary to make a small opening adjacent to the depression, through which a

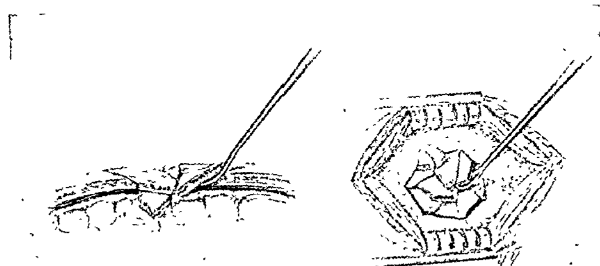


FIGURE 5. Sketch illustrating comminuted depressed fracture with underlying dural and cortical laceration. This type of fracture must be treated by elevating and removing the in-driven fragments rather than simply elevating the outer table into position.

curved dissecting instrument may be used to elevate the fragments into position. This can only be done if there is not extensive fragmentation of the inner surface of the skull. In the event that there are numerous depressed fragments, it will be advisable to remove these carefully with a rongeur. Large fragments with a periosteal attachment should be preserved. Any dural rent should be further opened and any necrotic brain tissue removed and a careful search made for any small, indriven bone fragments. If the dura has not been torn, it is advisable to make a small opening if there is any suggestion that subdural bleeding may be present. The dura should be closed. This may require a graft in some cases if there is extensive dural loss. This graft may be taken either from the temporalis fascia or if necessary from the fascia lata.

Fear of leaving a cranial defect should not interfere with the completeness of debridement of the small fragments which are separated from their pericranial attachments. Cranioplasty may easily be carried out either at this or at a subsequent time. Since there is no communication to the outside, elevation of a simple, depressed fracture may be deferred until such time as the patient's condition warrants proceeding with surgery.



FIGURE 6 (a) Compound depressed fracture of right frontal temporal region in a five-year-old boy showing large depressed bone plate with preservation of periosteal attachment.



FIGURE 6. (b) Post-operative photograph of 6(a) showing plate elevated and wired into position. Note silver clips at cribiform plate at site of dural tear and brain laceration. Temporal fascia used to repair dural laceration.



FIGURE 6 (c) Lateral view of 6(a) showing Comminuted Depressed Fracture. (d) Post-operative x-ray of same showing bone plate elevated and wired into position with restoration of skull contour

Compound Depressed Fractures

The fact that the depressed fracture is compounded to the outside [illegible] different management. Prompt [illegible] prevent the development of infection. This must, however, follow thorough evaluation of the patient for associated injuries, correction of shock, and the obtaining of proper x-ray views. Antibiotic therapy should be instituted promptly. Penicillin or one of the wide-spectrum antibiotics may be used.

At operation the scalp should be thoroughly shaved and cleansed with soap and water. Following this, thorough debridement and irrigation must be carried out using a separate tray. Careful attention should be paid to fashioning the incision in such a way that maximum exposure of the fracture site may be obtained and to the fact that rotation flaps may be necessary if the debridement necessitates removal of a significant amount of devitalized scalp.

Following debridement and cleansing of the soft tissues attention is directed to the fracture site. It must be re-emphasized that a relatively small depression of the outer table may overlie an extensive depression of the inner table. Particularly in compound fractures no attempt should be made to preserve small, detached pieces. One should not rely on levering up the area of depression from an adjacent burr hole since in this way major depressed fragments of the inner table may be overlooked. Frequently it is possible to start the debridement at one shelving edge of the depressed area by nibbling with a rongeur. This may afford sufficient space to begin gentle and careful dislodgement of wedged fragments with a small curved periosteal elevator or the rongeur. Great care should be taken to obtain sufficient space so that fragments can be lifted out with a minimum of twisting or manipulation. Lack of adequate care here may result in unnecessary dural laceration. This is of extreme importance when the area of depression is adjacent to or overlying a major venous sinus. Fragments which are depressed over or into a major sinus must be elevated and removed. All preparations, however, should be made to handle major venous hemorrhage. This presupposes that the edge of the calvarium surrounding the depressed fragment in the sinus has been rongeured away sufficiently so that exposure can be obtained promptly and hemorrhage from the sinus controlled without delay. Usually an oxycel pledget or a small piece of muscle held in place with moist cotton will control the hemorrhage. In rare instances it may be necessary to ligate the sinus, but should never be done in the region of or posterior to the rolandic inflow.

Del. Hosp. 254314

J. A., a nine-year-old white female, was admitted to the hospital on August 7, 1957 following an injury sustained in an automobile accident.

On examination in the emergency room the child was noted to be slightly stuporous but would respond coherently to questions and commands when aroused. There were two lacerations over the frontal scalp through one of which necrotic brain tissue was exuding. The pupils were equal and reacted to light. There was no choking of the disk or retinal hemorrhages. There was no facial weakness and the tongue protruded in the midline. The patient was able to move the upper extremities equally well. Neurologic examination of the lower extremities was interfered with by a fracture of the right femur. However, there was no paralysis evident.

X-ray examination revealed a badly comminuted and depressed fracture of the right frontal bone. The fracture of the right femur was also shown on x-ray. After emergency immobilization of the femur and correction of initial shock, the patient was taken to the operating room where



FIGURE 7. Late postoperative result following replacement and wiring of extensive comminuted depressed fragments of the frontal bone.

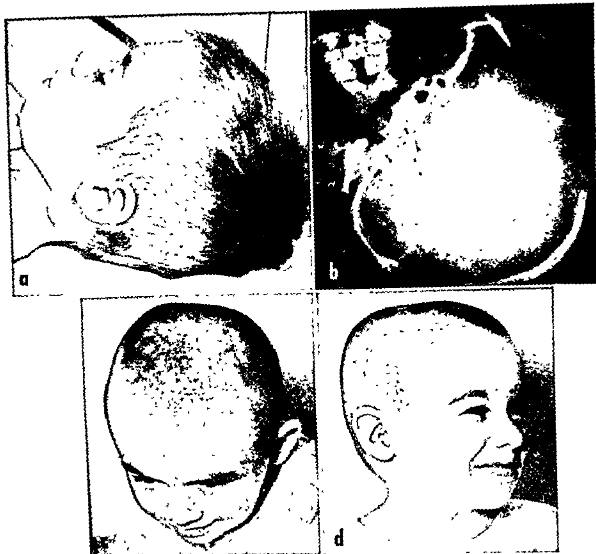


FIGURE 8. (a) Triangular file driven through frontal bone and into frontal lobes. (b) X-ray of same. (c) Postoperative photographs showing use of coronal incision and underlying right frontal flap to accomplish removal of foreign body without further brain laceration.

under general anesthesia, after the head was prepared and draped, a right frontal flap was made. The areas of depression in the right frontal bone had lacerated the dura producing a laceration of the medial portion of the right frontal lobe at the pole and extending along the base over the orbital plate. The dural incision was enlarged somewhat and the necrotic brain removed by suction dissection. After all necrotic brain and small bone fragments were removed, a temporal fascia graft was placed and sutured into position. The bone flap was then replaced and a two-layer scalp closure carried out. Plastic closure of the small frontal scalp lacerations was also carried out at this time.

Post-operatively the patient made an excellent recovery and showed no neurologic abnormality as a result of this injury. The fracture of the right femur was maintained in traction and subsequently the right leg was placed in a cast.

This case illustrates the situation in which a major procedure may be necessary in order to adequately cleanse and debride a brain and covering tissues and resort to dural grafting. These steps are essential for a proper outcome of the case.

Large fragments may be retained and either fitted into position or wired back if their pericranial attachment has been lost. Small, shattered fragments should be discarded. It is particularly important that all possible bone be saved in areas such as the glabella and supra-orbital ridge. Satisfactory cosmetic results from cranioplasty are most difficult to obtain in these areas. In compound wounds it is advisable to defer cranioplasty for three to six months even though there are reports of satisfactory results when the cranioplasty was done at the time of primary debridement.

Once the bony fragments have been satisfactorily attended to, any lacerated brain must be carefully debrided and a tight dural closure accomplished, either primarily or by use of a fascial graft. The scalp may then be closed, preferably in two layers if a satisfactory galeal layer has been preserved. A one layer closure with either silk or steel wire may be used if the galea is too friable to permit a complete two layer closure.



FIGURE 9. Bone defect resulting from skull fracture in infancy with production of lepto-meningeal cyst.

Linear Skull Fractures—Simple and Compound

As every pediatrician and neurosurgeon is aware, the prospect of a skull fracture seems to arouse more terror in the minds of the parents than what has actually happened to the brain. The demonstration of a simple, linear or comminuted, non-depressed fracture on the x-ray has no significance so far as treatment is concerned and indicates only that a severe blow has been sustained. If such a linear or comminuted fracture underlies a scalp laceration, then careful attention must be given to the most meticulous technique of debridement and closure of the scalp laceration in order to avoid any opportunity for the development of infection.

In young children one may see the development of a leptomeningeal cyst. This results from a linear fracture with an associated dural tear. An arachnoidal cyst develops. As the cerebral pulsations continue the fracture line tends to widen by bone resorption at the edges. A very wide interruption in cranial continuity may result. Surgical correction of this lesion is necessary. The incision should be placed so that it does not immediately overlie the fracture site. The dural tear must be carefully delineated and a fascial graft used to cover the defect. Cranioplasty is not usually necessary at least in the very young child since the skull defect will tend to close. In the child of five years or older with a significant defect, cranioplasty with autogenous bone, plastic, tantalum, or steel is advisable.

Rhinorrhea and Otorrhea

These two conditions are not uncommonly seen with fractures of the base of the skull or fractures extending into the paranasal sinuses. Cerebro-spinal rhinorrhea results from a fracture line extending into the frontal sinus or across the cribiform plate. Associated with the fracture line there is, of course, a dural tear. The cerebro-spinal fluid has a method of egress from the subarachnoid space into either the frontal or ethmoidal sinus or directly into the base of the nose. Careful observation should always be carried out in order to determine whether or not this finding is present. Should there be any nasal bleeding this should be examined to see whether or not there is any admixture of cerebro-spinal fluid.

Otorrhea, or drainage of cerebro-spinal fluid from the ear, is frequently seen with basilar fractures involving the petrous ridge or with fractures of the vault in which the fracture line extends down into the petrous ridge. The drainage of thin, bloody fluid from the ear is indicative of such a fracture. Routine x-rays of the skull will often not reveal the fracture line and special views of the base may be necessary, when the patient's condition permits, in order to attempt to demonstrate the fracture line.

This condition is, of course, a very serious complication of skull fracture. It brings with it the danger of meningitis. Fortunately, with modern anti-

made to completely tabulate and describe all of the various materials. The most commonly used materials and techniques for cranioplasty are as follows:

1. *Osteo-periosteal grafts:* This type of cranioplasty is particularly suitable for the relatively small defect in young children. In a child who is old enough to have well formed inner and outer tables and diploe, the parietal eminence is selected as a donor site. The site to be grafted is first carefully prepared. If the scars of previous wounds or surgery permit the incision is made away from the graft site. Care is taken not to incise or destroy the pericranium. After the scalp is carefully dissected free, the pericranium is incised about the margins of the bony defect. The scarred pericranium together with the scar overlying the dura is then dissected away centrally. Care is taken not to penetrate the dura. If the dura is defective, the meningo-cerebral cicatrix should be gently separated and either temporal fascia or fascia lata used to close the dural defect. The final step in preparation is to freshen the bone edges with a rongeur and remove any projecting spicules.

Attention is then directed to removing the graft. A curved or linear incision is made over the parietal eminence. A pattern the size of the defect is then placed over the pericranium and the pericranium is incised around the margins of this pattern. Minimal separation of the pericranium at the margins is then carried out. The outer table is then penetrated with a chisel or osteotome or a large trephine may be used. In this fashion the bone plate is separated and removed. Care must be exercised in order not to penetrate the inner table. After the graft has been removed, the intact pericranium is used to suture the graft into place in the recipient site. Routine scalp closure is then carried out.

2. Other *autogenous bone* may be used as desired. The *iliac crest* may be used to cover somewhat larger areas and may give a very satisfactory skull contour. *Split rib grafts* may make a very satisfactory type of bone for the repair of skull defects. The use of a rib may particularly be helpful in fashioning a supraorbital ridge. Wire sutures placed at the margins of these grafts will satisfactorily secure the bone in position.

3. The most promising *plastic cranioplasty* material is methyl-methacrylate. The use of this material as well as a very simple and satisfactory technique has been developed by Spence. The method is useful because of the rapidity and simplicity with which large complex plates may be fashioned at the operating table. The fixation is simple and the resulting skull contour excellent. The same principles apply as with any other method of closing a cranial defect. The scalp incision should not be placed over the cranial defect if it can be avoided. The bed must be properly prepared by excision of scar and freshening of the bone edge so that the plate may



FIGURE 12. Postoperative skull x-ray showing area covered by form-fitting cranioplastic plate secured by wire sutures. The plastic material is nonradiopaque.

be bedded properly. Screws or wires may then be used to fasten the plate into place. Multiple drill holes should be made through the center of the plate to prevent fluid collection beneath the plate and to permit the growth of fibrous tissue through the plate for better fixation. Fluid accumulation beneath the scalp may require several aspirations.

As a result of extensive experience with this material, Spence feels that one can form such a plastic plate for use in young children even though bone growth is still going on.

4. *The use of metallic plates: Tantalum or stainless steel plates* are of great value in covering cranial defects in either adults or children. The techniques of formation and fixation of these plates requires meticulous attention to detail. Proper preparation of the area is essential. The plate must be an exact fit. This may be accomplished by making a positive cast of the defect at the original operation and forming a negative from this. The plate may then be formed under high pressure in a closed container. Holes are then drilled in the plate to prevent the trapping of fluid. Minor

adjustments may be needed at the operating table before fixation is completed. The plate may also be formed at the operating table by the use of various positive and negative moulds. Several manufacturers also supply standard preformed plates which may require only minor manipulation in the operating room. The use of metallic plates may necessitate a somewhat longer time taken at the operating table. It is best not to insert a metallic plate in the presence of actual or potential infection. Three to six months should be allowed to elapse before re-operation and the insertion of a metallic plate.

Sequelae of Acute Head Injuries in Children

Cranial Nerve Lesions

Various cranial nerve palsies may be seen following head injury. Probably the cranial nerve palsy most frequently seen involves the first or olfactory nerve. In a child, this may easily be overlooked. Severe trauma to the head with shifting of the basal portion of the brain may disrupt the fine terminations of the olfactory nerve with resulting unilateral or bilateral anosmia.

Direct damage to the optic nerve may occur either from a fracture in the region of the optic canal or from a shearing stress placed on the optic nerve resulting from severe trauma. This lesion is not common but careful evaluation, nevertheless, should be carried out in order to avoid overlooking a major neurologic deficit.

A third nerve palsy is frequently encountered. This may be complete or incomplete. Probably the most common cause is herniation of the medial portion of the temporal lobe over the tentorial edge with pressure on the third nerve. This should eventually clear following correction of the compressing clot. The third nerve may also be involved, however, on the basis of direct contusion or sudden stretching of the nerve. Ptosis, a dilated and fixed pupil and impairment of medial rotation of the eye will, of course, result. Careful observation and acute clinical judgment will be necessary in the initial stages in order to differentiate this from a third nerve palsy secondary to a compressing intracranial clot. The prognosis for recovery of third nerve palsy on the basis of contusion or stretching is usually good.

A sixth nerve palsy is a not uncommon finding and may result from increased intracranial pressure as in a subdural hematoma or subdural hygroma or it may result from direct trauma to the nerve. The sixth nerve has the longest course of any cranial nerve beneath the brain stem and is therefore subjected to stresses as a result of sudden shifting of the brain

or displacement from increased intracranial pressure. When secondary to the presence of a space occupying clot, the palsy will clear following removal of the compressing clot. When the sixth nerve paralysis is secondary to sudden stretching of the nerve, the prognosis is usually good but clearing of the lesion may take several weeks or months.

The seventh and eighth nerves may be involved together in a fracture



FIGURE 13. Five-year-old boy who sustained a left posterior temporal skull fracture and left sixth nerve palsy. Sixth nerve palsy secondary to direct nerve injury rather than increased intracranial pressure. Patient looking to left.

of the petrous ridge. The facial palsy may be immediately evident if the injury is severe with direct trauma to the facial nerve. The appearance of facial paralysis may be delayed somewhat and appear only after several days. In this instance, it is usually secondary to edema of the nerve within the facial canal. The prognosis for eventual recovery is somewhat better than if the palsy is immediate in onset. The presence of a facial palsy with or without an obvious fracture of the petrous or mastoid bone should lead to a careful hearing examination. The child may not realize that there is any impairment. Careful examination will demonstrate that hearing impairment is not frequent with fractures involving the middle fossa and petrous ridge.

Involvement of the ninth, tenth, eleventh, and twelfth cranial nerves

usually implies an injury of such severity that it would be incompatible with life.

Focal Cerebral Deficits

Cerebral contusion or laceration, with or without intracerebral hemorrhage, may be responsible for lasting focal neurologic deficits. The clinical picture will, of course, depend on the area involved. Aphasic speech disorders may be seen with focal damage either in the left inferior frontal region or angular gyrus region. The prognosis for recovery is, of course, much better in a child than in an adult, particularly with prompt and effective speech training.

Motor deficits of varying degree may be seen with injuries in or adjacent to the motor cortex or its projection pathways. Unless severe and extensive destruction has occurred, partial or complete improvement is the usual course. Weakness of the hand and arm may occur early in the course of the illness, suggesting the presence of a compressing clot. Usually, however, focal damage of this type is not accompanied by general deterioration of the state of consciousness. However, it may, of course, in any single in-

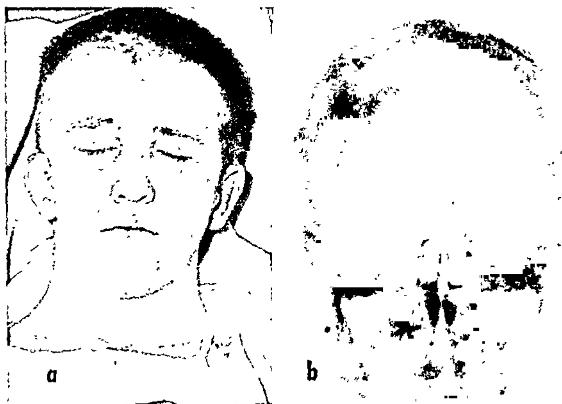


FIGURE 14. (a) and (b) Photograph and x-ray reproduction revealing severe compound comminuted depressed fracture frontal bone necessitating removal of many small comminuted fragments. Extensive frontal lobe damage with longitudinal sinus thrombosis Improvement but with severe intellectual and personality defect and residual right hemiparesis.

stance, be necessary to perform multiple trephine exploration in order to rule out a space-occupying clot. Recovery may be hastened by starting physiotherapy as early in the post-traumatic period as the patient's condition will permit.

Severe frontal lobe injury may result in defects in memory, intellectual power, and personality changes of varying degree. With compound depressed fractures of the frontal and/or parietal areas in the midline with extensive damage, thrombosis of the longitudinal sinus may result with a characteristic picture of tetraplegia or tetraparesis. With the less severe degrees of damage, the course is usually one of progressive improvement. Some degree of personality defect and intellectual impairment may, however, remain. Psychometric evaluation after the acute effects of the trauma have cleared is important in assessing this damage and attempting to prognosticate for the future.

One may occasionally see severe post-traumatic psychotic pictures after severe generalized cerebral contusion in children. There may be a total personality deterioration with wild, restless, and combative behavior. Though the picture initially may be suggestive of a poor eventual prognosis, usually there is progressive clearing with lessening of the agitation, more rational behavior and return to a more normal personality pattern.

Post-Traumatic Epilepsy

Post-traumatic epilepsy may be seen as a sequela of either closed or open head injuries. The incidence is low in closed trauma and the figures vary from 2 to 6% of cases reported by various authors. Seizures may begin in the early post-traumatic period or may be delayed for weeks or months. The likelihood of post-traumatic seizures coming on after a period of two years is, according to Walker (1949), unlikely. Post-traumatic seizures may be either focal or generalized in character.

The frequency of seizure occurrence rises sharply in the open head injuries with brain laceration and contusion. Areas of focal damage serve as irritant foci with the production of seizures. Anticonvulsant medication should be promptly instituted following the appearance of seizures. We have not followed the practice of prescribing anticonvulsant medication routinely in the absence of any seizures though in some clinics this is felt to be advisable. Focal areas of cerebral damage demonstrated by focal spiking in the electro-encephalogram or by alterations in the air-encephalogram may, when seizures are intractable on adequate medication, require later excision. It is, of course, self-evident that the proper handling of head trauma in the acute stages will minimize the incidence of this distressing post-traumatic complication. Depressed fractures with bony spicules projecting inward toward the cortex are of particular importance in this regard.

Cerebral Atrophy

One of the late sequelae of severe generalized head trauma is cerebral or cortical atrophy. This atrophy may be local or it may be generalized and diffuse. This pathologic process is usually disclosed in the course of complete investigation to determine the cause of post-traumatic seizures or focal neurologic deficits appearing after trauma. This process indicates a severe degree of brain damage and the prognosis is poor for recovery in view of the structural change. If the atrophy is focal, and results in intractable seizures, then surgical excision monitored by electro-corticography is indicated.

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CHAPTER XVI

Trauma—Subdural Effusions and Hematomas in Infants

ROBERT F. MABON

SUBDURAL hematomas and effusions in infants under the age of two years are quite common in occurrence. However, only during the past decade have enough cases been assembled to yield the necessary information obtained by close scrutiny of a large series for an intelligent approach to the diagnosis and management of patients with this lesion. The marshalling of this data, as well as an aroused interest in this entity, have provided the pediatrician and neurosurgeon with the information required to treat successfully and care for a condition which has a very satisfactory outcome if properly managed.

Incidence

The greatest incidence of subdural hematomas and effusions in infants occurs around six months of age. Most clinics have found an increase in the number of patients with this lesion in recent years. This undoubtedly is due to the alerted attitude of physicians particularly interested in the treatment of diseases of children. Guthkelch recently stated that of all the surgical conditions of the central nervous system occurring in the first year of life, only spina bifida and hydrocephalus were seen more often than the subdural hematoma. Subdural hematomas are reported by Sulamaa and Vara to be ten times as frequent in breech presentations as in spontaneous deliveries. They also found this lesion about twice as often in children of primaparae as in multiparae.

Etiology and Pathogenesis

Trauma is the most common etiological factor in subdural hematomas and effusions in infants. Most frequently the injury occurs at birth and symptoms are not evident until sometime later during the first year of life. Accidents as being struck on the head or falling out of bed actually make up a very small percentage of such patients in this age group. The

site of the bleeding is usually the bridging veins as they traverse the dura coming from the cerebral cortex to their termination in the superior sagittal sinus. These vessels are particularly vulnerable to shearing forces resulting from the moulding of the parietal bones as the infant passes through the birth canal. A protracted, difficult labor will result in more frequent tearing of these fragile vessels than in an easy delivery. In the normal uncomplicated delivery, prolonged labor may well play an important etiological role. These very pertinent observations underline the findings of Chase who demonstrated the close relationship of tentorial "splits" and subdural hemorrhage in infants. He also considered prematurity with concomitantly less well developed protective intracranial membranes, e.g., the dura mater, as an important causative agent.

In instances with no definite cogent predisposing agent, it is entirely possible that the previously mentioned causes, i.e., prematurity, birth injury, difficult labor, may be the causative agent if pertinent historical data were available. The more one encounters this condition, the greater the conviction that adequate statistical information will affirm the clinical impression that trauma of one sort or another is the major precipitating factor in the great majority of instances. Elvidge and Jackson, and Jackson and Werner, have emphasized that the varying states of malnutrition, vitamin deficiency, systemic infectious processes, etc. may aggravate the already existing clinical picture but they are not in themselves responsible for the hemorrhage. These accompanying enervating states may be the result of the hematoma rather than the cause. Examples of this syndrome which are secondary to postnatal trauma have been discussed under the general subject of head injuries.

Other agents have been considered as predisposing factors if not primary causes. Various enfeebling systemic conditions, i.e., blood dyscrasias, vitamin C and K deficiencies, infectious processes located elsewhere in the body, and malnutrition have been quoted as precipitating agents. None of these are consistent and uniform enough in their appearance to consider as major causes.

Rarely one will encounter subdural collections of colorless fluid, commonly classified as subdural hygromas or hydromas. These cases have been described by Dandy as probably resulting from traumatic focal tears or rupture of the arachnoidal membrane, with subsequent escape of the cerebrospinal fluid into the closely adjoining subdural space. A ball valve-like action may result with the fluid escaping rather easily through the rent in the arachnoid into the space beneath the dura, but with relatively little chance for the fluid to re-enter the subarachnoid space. The fluid transfer actually may be due entirely to a mechanical defect or to the fact that whatever caused the traumatic rupture, also injured the general

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Etiology and Pathogenesis

Trauma is the most common etiological factor in subdural hematomas and effusions in infants. Most frequently the injury occurs at birth and symptoms are not evident until sometime later during the first year of life. Accidents as being struck on the head or falling out of bed actually make up a very small percentage of such patients in this age group. The

site of the bleeding is usually the bridging veins as they traverse the dura coming from the cerebral cortex to their termination in the superior sagittal sinus. These vessels are particularly vulnerable to shearing forces resulting from the moulding of the parietal bones as the infant passes through the birth canal. A protracted, difficult labor will result in more frequent tearing of these fragile vessels than in an easy delivery. In the normal uncomplicated delivery, prolonged labor may well play an important etiological role. These very pertinent observations underline the findings of Chase who demonstrated the close relationship of tentorial "splits" and subdural hemorrhage in infants. He also considered prematurity with concomitantly less well developed protective intracranial membranes, e.g., the dura mater, as an important causative agent.

In instances with no definite cogent predisposing agent, it is entirely possible that the previously mentioned causes, i.e., prematurity, birth injury, difficult labor, may be the causative agent if pertinent historical data were available. The more one encounters this condition, the greater the conviction that adequate statistical information will affirm the clinical impression that trauma of one sort or another is the major precipitating factor in the great majority of instances. Elvidge and Jackson, and Jackson and Werner, have emphasized that the varying states of malnutrition, vitamin deficiency, systemic infectious processes, etc. may aggravate the already existing clinical picture but they are not in themselves responsible for the hemorrhage. These accompanying enervating states may be the result of the hematoma rather than the cause. Examples of this syndrome which are secondary to postnatal trauma have been discussed under the general subject of head injuries.

Other agents have been considered as predisposing factors if not primary causes. Various enfeebling systemic conditions, i.e., blood dyscrasias, vitamin C and K deficiencies, infectious processes located elsewhere in the body, and malnutrition have been quoted as precipitating agents. None of these are consistent and uniform enough in their appearance to consider as major causes.

Rarely one will encounter subdural collections of colorless fluid, commonly classified as subdural hygromas or hydromas. These cases have been described by Dandy as probably resulting from traumatic focal tears or rupture of the arachnoidal membrane, with subsequent escape of the cerebrospinal fluid into the closely adjoining subdural space. A ball valve-like action may result with the fluid escaping rather easily through the rent in the arachnoid into the space beneath the dura, but with relatively little chance for the fluid to re-enter the subarachnoid space. The fluid transfer actually may be due entirely to a mechanical defect or to the fact that whatever caused the traumatic rupture, also injured the general

intracranial contents to the point where a considerable amount of edema resulted increasing the cerebrospinal fluid secretory processes.

Later the aperture may seal over completely, thus preventing any reabsorption of the entrapped fluid. The membranes encapsulating this type of effusion may be exceedingly tenuous or non-existent in some instances, because the irritating stimulus which activates the cellular proliferation in the hemorrhagic effusion, i.e., the catabolic decomposition products of whole blood, is lacking. In many cases this lesion is easily correctible by trephination and drainage alone, although occasionally one will encounter a more recalcitrant lesion of the sort that may necessitate secondary trephine drainage or even an osteoplastic craniotomy.

Whatever the etiological agent, bleeding in various amounts occurs in the subdural space. Initially, the clot is surrounded on the outermost aspect by the mesothelial cells of the dura, and on the inner surface by the cells composing the arachnoid membrane. The cells of the inner boundary of the dura, being more primitive in origin (mesoblastic), are thought to react more quickly to foreign stimuli, and for this reason will proliferate more rapidly than the more highly differentiated cells of the arachnoid, whose origin is from the neural crest. Thus, in the initial phases, the growth is more extensive along the outer margins of the clot, and when fully developed this membrane, the so-called neomembrane will be thicker than the corresponding more filamentous innermost structure formed by the cells comprising the delicate arachnoid layer. In the early stages, the neomembrane strips reluctantly from the dura, and any attempt to do so will result in brisk bleeding. Later in the course of the disease, it can be more readily separated. The entire picture of fibroblasts surrounding and growing into the clot is a natural attempt to granulate the foreign body, i.e., the clot. This is seldom successful if the original hemorrhage is of any size because the ground work is laid for the old experimental set-up, of a semipermeable membrane lying between fluids of greatly differing osmotic pressure. The constituents composing the clot will break down into fractionated elements, primarily the proteins contained in the encapsulated blood and form a fluid exerting a greater osmotic pressure than the closely adjacent cerebrospinal fluid. This will, of course, give rise to a flow of the less dense into the fluid of greater density, and in effect swelling the contents of the clot to one of considerably greater bulk than the original dimensions. Therefore, without any further bleeding, the initial hemorrhage can enlarge slowly and relentlessly to the point where the intracranial pressure becomes very high.

The Clinical Picture

There is no consistent clinical pattern of infants with this condition. It should always be entertained as a differential diagnosis in any infant with

suspected hydrocephalus. In a number of cases, subdural hematomas will be encountered in the course of a routine work-up of hydrocephalic infants. This pertinent observation should be underlined repeatedly to those interested in pediatrics or the general practitioner who may carelessly dismiss a child as a "hopeless hydrocephalic." Even this diagnosis and attitude can-



FIGURE 1. Photograph of infant with subdural effusions showing the slight enlargement of head with fullness of the anterior fontanelle. (*Courtesy of* Dr. Ira Jackson)

not be condoned, but certainly a mistaken interpretation of this sort could be tragic. The accelerated head size is ordinarily not nearly as pronounced as in other types of hydrocephalic infants. The association of increased head size with a bulging, taut fontanelle is very significant. Other than the obvious hydrocephalic, there are additional presenting features which make one suspicious of this syndrome. The most commonly encountered symptoms and signs are incorporated in Figure 2. In any infant presenting a bulging, tense, anterior fontanelle, with or without fever, one should immediately consider the presence of a subdural effusion. Vomiting usually occurs immediately after an ingested meal, and it may be projectile but not invariably so. This is a real problem because as has been indicated, malnutrition is a distressing complication and can be compounded by persistent regurgitation. Convulsions are rather common. Seizures may vary from focal twitchings of the face to generalized attacks. Intensive anticonvulsive measures may be required because prolonged attacks profoundly weaken an infant who is already in an exhausted and generally

trated in the chapter on diagnostic techniques. Under normal conditions no fluid is obtained from the subdural space. If several drops or even a cubic centimeter or more of colorless fluid appear, one may have introduced the needle too deeply and is actually in contact with cerebrospinal fluid in a large subarachnoid cistern. A similar experience may also occur in porencephaly with a large loculation of cerebrospinal fluid, not communicating with the usual cerebrospinal fluid channels and pathways. Occasionally in the presence of an unsuspected, tremendous hydrocephalus, one will encounter a cerebral cortex so thin that the needle may penetrate the tenuous cortical layer almost as soon as the fontanelle membrane is pierced. In such instances one must not be misled because on occasion the ventricular fluid may be deeply xanthochromic with an obviously elevated protein content. Infrequently, one may have the experience of puncturing a large vein, but this error is readily recognized since the blood will flow sluggishly and clot quickly. In this instance, the needle should immediately be withdrawn. In subdural hematomas, particularly in relatively early cases, the fluid is usually grossly bloody but definitely xanthochromic. At times, the xanthochromia may be very faint. It is always unwise to attempt aspiration for fear of injuring the cerebral cortex. The encapsulated fluid will flow quite readily especially through a No. 18 needle and even spurt rather forcibly in some instances. A bilateral tap is mandatory because about 75% of these patients have bilateral subdural effusions. For the purpose of diagnosis enough fluid is removed to relieve the increased pressure. The removal of large amounts may precipitate alarming systemic symptoms as will be discussed under treatment.

Pneumoencephalography and Ventriculography

Neither of these procedures are absolutely imperative to establish a diagnosis so readily and accurately made by subdural puncture. They are most useful during the postoperative period when the patient is not responding too well to what has apparently been adequate therapy. Ventriculography is usually not necessary except when the intracranial pressure is elevated and there is uncertainty about the diagnosis. The complication of a communicating internal hydrocephalus may in this way be demonstrated. The pressure of the effusion may compress the normal subarachnoid spaces over the cerebral hemisphere to such an extent that there is interference with the normal absorptive mechanism. Injection of air by the spinal route will be more informative in the presence of a nontense anterior fontanelle and in the absence of other signs of increased intracranial tension, because of better visualization of the subarachnoid spaces An abnormally large accumulation of air in this compartment, as well as an enlarged ventricular system may indicate that irreversible changes have taken place as a result of the lesion. This knowledge can be

extremely helpful in evaluating the future development of the patient. If the compression of the subarachnoid spaces is not pronounced, or if a moderate degree of cerebral atrophy exists, the pneumoencephalogram will reveal air abnormally distributed throughout the subarachnoid spaces. Roentgenograms may be made following partial replacement of the aspirated subdural fluid with air but ordinarily this is not necessary.

Treatment

The final objective in infants with subdural hematomas or effusions is to completely remove the fluid and obliterate the subdural space, thus restoring the normal compartmentation of the intracranial structures. As long as there remains a dead space in the subdural cavity, further accumulation of fluid will occur.

Aspiration

Aspiration alone will serve not only to confirm the diagnosis but may also be an important therapeutic measure. In early cases, when the condition has existed for only several weeks to a month, rarely a single aspiration, and in some instances repeated punctures, may effect a cure because no neomembrane has formed as yet. However, it is well to emphasize that this is also a valuable procedure in situations where there is no question as to the size or age of the effusion, and the fluid is definitely xanthochromic. In the latter event, unquestionably the condition has existed long enough to establish sturdy neomembranes. On the other hand, one should be wary of the formation of the neomembrane in those instances in which a relatively early effusion has been seemingly adequately treated. Trephination or pneumoencephalography may be necessary in these instances to adequately rule out this complication to avoid the possibility of compromising the future growth and development of the cerebrum. In long standing cases, with the patient in a critical state, a subdural puncture may actually be a life-saving procedure. It is best in instances of this sort to remove just enough fluid to permit a reduction of the intracranial hypertension. The removal of large quantities of fluid alone may precipitate alarming symptoms due to the sudden alteration in the intracranial fluid-pressure relationship. Doing so can produce profound shock with the infant in an obviously critical state, virtually pulseless, breathing quite rapidly and having the ashen grey facies characteristic of a shocked state. It may be necessary to repeat the puncture daily in some patients to tide them over until a more direct attack is feasible. During this period the infant's general condition should be improved. Whole blood may be administered intravenously to combat any anemia and other general measures taken to prepare the child for more definitive procedures such as craniotomy.

In moderately acute situations, the fluid aspirated may be grossly bloody.

As the taps are repeated, the fluid will tend to be less sanguinous and more xanthochromic in appearance. Elvidge and Jackson have shown that with daily aspiration an infant may lose up to 5% of his daily protein need and 10% of his daily fluid requirement. There is a constant siphoning of the body proteins and fluids into the subdural effusion, and repeated aspirations increase this loss. Therefore a vigorous effort should be made to replace the protein and fluid loss.

Trephination

After the diagnosis has been made by subdural taps, the patient is taken to the operating room where burr holes are done. Burr holes are advised in these patients for several reasons. The main concern is the presence or absence of a membrane (Figure 4b). The burr holes are placed in the temporal parietal region so that they can be incorporated in the bone flap if a craniotomy is done. These are done bilaterally and Penrose drains may be inserted in the subdural space. When no membrane is found the drainage may continue as long as five days. If a membrane is present the drainage will continue as long as the drain or membrane are present. Therefore in such cases craniotomy should not be delayed too long. Burr holes are done under general anesthesia with a cut down in the ankle vein to insure adequate fluids and blood. The dressings should be changed daily and one must constantly protect the wounds from infection. Very often the exploratory trephine openings can be done in conjunction with and prior to a contemplated craniotomy.

The purpose and ultimate objection in the treatment is to obliterate the existing dead space.

Craniotomy

When there is a sizeable clot, with an organized hematoma or an inner membrane, craniotomy is imperative. However, the operation should be performed only when the child is in the best possible condition. Every preoperative precaution to insure the success of the operation should be utilized. A preoperative phlebotomy or "cut down" to insure proper fluid and blood replacement is imperative. The accessability of the terminal portion of the great saphenous vein, lying midway between the anterior border of the medial malleolus and the tendon of the tibialis anticus, makes this vessel the choice of many. A procedure of this magnitude should not be initiated without this important preliminary step. Any infant tolerates rather poorly blood and fluid loss during surgery, since only a relatively small per cent depletion may mean the difference between a catastrophic and a highly successful operation. Fluids and whole blood must be given during the operative and postoperative periods. The high protein content in



FIGURE 4. (a) Photograph at surgery of a burr hole with the dura opened at revealing the underlying tense outer membrane of the subdural effusion. (b) Photograph after the outer membrane has been removed at the burr hole site along with some of the subdural fluid revealing the underlying brain at a distance away from the dura. (c) Photograph identifying the constricting inner membrane through the burr hole. (*Courtesy of* Dr. Ira Jackson)

the hematoma may have an impoverishing effect on the general body protein content, particularly when one is dealing with a chronic hematoma. The infant customarily is critically ill and as a result may not be taking the proper amount of needed protein by diet, due to poor appetite, vomiting, etc., and will need a greater reserve because of his generally malnourished state. Fever and other enervating factors may further ravish this reserve stock.

The craniotomy, which is a fronto-temporo-parietal osteoplastic or free bone flap, should be quite large so as to adequately visualize both frontal and occipital poles and to be able to remove the neomembranes in their entirety, since the hematoma may be quite large in its final development. As has been mentioned earlier the original hemorrhage is frequently small but because of the breakdown of the blood components of the original hemorrhage, mainly the blood cells, the protein increase which results from this catabolism will attract the cerebro-spinal fluid into the subdural compartment enlarging the original effusion many times its original size. The craniotomy very often can be accelerated by using scissors to section the bone between the burr holes. Some surgeons prefer a Gigli saw, but this is a matter of personal preference. It is unusually difficult if not impossible to separate the dura from the underlying thickened outer neomembrane. The two most often are reflected as a unit and later can be more readily separated. When an incision is made in the dura, there may be a gush of xanthochromic fluid and a sudden decompression with collapse of the bulging membranes. The clot may be solid in some instances, or composed of mingled clot and fluid and should always be completely removed. The inner membrane may be filmy to very tough.

Usually, the thin inelastic membrane can be teased away from the pia-arachnoid overlying the cerebral cortex as Figure 4c. It should be completely excised from the frontal to the occipital poles. The removal of the membrane may be done by either blunt or sharp dissection. One, however, will find in many instances small islands of the inner membrane which may be extremely adherent to the arachnoid and underlying cortex. Not only may brisk, difficultly controlled bleeding result from dissection of these islands but actual harm may be done to the cortex itself by persisting in this dissection. Removal of the major portion of the membrane will usually be all that is necessary. One should always have a good clear exposure as this is done.

An additional reason for a large craniotomy flap is because particularly toward the vertex one may tear bridging pial veins, which can produce an extremely annoying hemorrhage. The wound should be closed in the usual fashion. If the brain is adequately expanded to fill the space formed by the effusion it will not be necessary to drain the wound, although some sur-



FIGURE 5. (a) Photograph of the outer membrane with the dura reflected upwards at the time of the craniotomy. (b) Photograph showing the constricting inner membrane overlying the cortex with some subdural fluid. The outer membrane has been removed and the dural flap is turned upwards. (c) Photograph showing the removal of the inner membrane. (d) Photograph of the brain after the inner membrane has been completely removed. Note the extent of the subdural space indicating lack of expansion of the brain. (e) The membranes have been removed and the brain expanded by means of injecting fluid into the lumbar subarachnoid space thereby completely obliterating the subdural space.

(*Courtesy of* Dr. Ira Jackson)

geons do. In some instances where cerebral expansion does not occur despite adequate compensatory measures as discussed below, the wound will require drainage. In these instances, however, one should be aware of the fact that prolonged drainage over twenty-four hours is somewhat hazardous for fear of a resulting infection. The drain is carried from an inferior position of the subdural space out through a separate stab wound opening in the scalp. In a patient with a bilateral effusion there is a delay of about one week before attempting the second stage. One utilizes this period by checking the state of the infant's general blood picture and again administering whole blood or whatever measures may be necessary to fortify the child for the second stage.

At times the brain may fail to expand sufficiently as shown in Figure 4d to fill the space remaining after removal of the clot or fluid. Elvidge and Jackson have facilitated expansion by injecting Ringer's solution into the lumbar subarachnoid space. Jackson and Werner have suggested doing this routinely at the beginning of operation. A lumbar needle is inserted and attached by means of a sterile tube to a bottle of solution held higher than the head. After the effusion has been evacuated the Ringer's solution is allowed to flow into the lumbar subarachnoid space by gravity. They emphasized the fact that the fluid should never be forced into the lumbar canal.

Ventricular injections may also be done for the same purpose but fluid leakage through the openings made by the cannula may nullify the results and not be as effective as the spinal route. In a few instances the process has existed so long it has caused pronounced gliosis of the brain so that expansion will not occur with any of the measures mentioned above. Jackson and Snodgrass have had several such patients who invariably proved to be mentally retarded. In these patients they performed a subdural peritoneal shunt without ultimate success. Of course, in very chronic cases one may not actually be dealing with failure of expansion alone. One must bear in mind that the head circumference may be larger in the individual instance if the lesion had not been present. Then one is dealing with a skull larger than an essentially normal developing intracranial contents, the remainder of the increased volume having been caused by and composed of hematoma. Consequently, one cannot always hope to obtain complete filling of the evacuated space after complete removal of the effusion and its limiting membranes alone.

Subdural Effusions Complicating Infectious Processes

Subdural effusions following bacterial meningitis unrelated to trauma or birth injury were first described in the literature as recently as 1950. Prior to the introduction of the newer chemotherapeutic and antibiotic substan-

ces, the morbidity and mortality rate of certain bacterial meningitic infections was so high as to preclude any but a very few children surviving this type of illness. However, in recent years, since the mortality rate has been lowered so spectacularly, one encounters subdural effusions in these patients with ever increasing frequency. The age group most commonly affected is from birth to one year of life. To a lesser degree, the number of patients with complicating subdural effusions accompanying aural and nasal purulent infections is also increasing similarly because of the increased use and success of the newer therapeutic preparations. Apparently, the incidence of subdural effusions is far greater in cases in which the infecting organism has been Hemophilus influenzae (Type B). Pneumococcus and meningococcus infections are next in frequency.

One should be alert to the possible presence of a subdural collection of fluid where the infecting agent causing a leptomeningitis is H influenzae. Regardless of the organism a subdural tap should be done if the child still appears ill despite what is seemingly an adequate amount of antibiotics with a bacteriologically negative spinal fluid and a diminishing spinal fluid cell count particularly if the anterior fontanelle is still tense. The presence of fever should not deter one from tapping these infants as many will have elevated temperatures. As in post-traumatic subdural collections of blood, the suspected diagnosis is easily confirmed by subdural puncture. The fluid may be blood tinged, xanthochromic, clear or, as is often the case, cloudy. Frequently one may recover the infecting organism from the fluid, but this is the exception rather than the rule. The protein content is usually elevated. Very often the lesion is bilateral. The surgical technique is essentially the same as described on pages 431-436.

Subdural aspiration may be all that is necessary but in many instances trephination with drainage or osteoplastic craniotomy, to permit removal of tough inelastic neomembranes, may be required. It is best not to carry out any surgical procedures until the patient is free of fever. Repeated subdural taps should be done until the patient is afebrile. Antibiotic therapy as given for the meningitis should be continued throughout the post-operative course. This type of subdural effusion is also discussed in Chapter X on Inflammatory Diseases.

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CHAPTER XVII

Trauma—Spinal Column

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TRAUMA of the spinal column may occur in a child of any age. It has been most frequently encountered in early infancy from circumstances incident to delivery and among teenagers when the expanding physical and social activities of these older children have exposed them to serious falls, athletic mishaps and vehicle accidents.

Birth Injury

The cervical portion of the spinal cord may be stretched or deformed during a delivery in which the mechanics are unusually difficult. Neurologic deficits often occur in such instances without roentgenographic evidence of subluxation, fracture or dislocation of the cervical vertebrae.

It is possible that some mild spinal injuries which are incurred at birth remain undetected and that some serious spinal injuries are masked by concomitant craniocerebral damage. When there is clinical evidence of serious spinal cord destruction, the prognosis for long-term survival is poor.

The neck should be maintained in a neutral position when the injury is acute. This may be accomplished by placing a folded towel beneath the curve of the neck and sandbags on each side of the head when the infant is in a supine position. The head and neck should be supported with a pillow when the patient is turned on one side. Some active infants with a severe spinal cord injury may require direct skull traction in order to maintain a satisfactory spinal posture. Immobilization of the neck is usually required for about two weeks. Gavage feedings are sometimes necessary. The same thoughtful nursing care is required for a newborn infant with a quadraplegia as in the case of an older child with the same affliction.

Sprain

A simple sprain of the vertebral column is the most common spinal injury encountered in infancy and childhood. Many patients who have suffered a head injury may show some degree of neck or lower back sprain. These combined head and spine injuries occur most frequently as a result of falls or vehicle accidents. When two vehicles collide, or one vehicle collides with

an immobile object, the spinal column of a victim may be violently flexed or extended in such a manner as to damage the supporting ligaments of both the lower cervical and lumbar spine without causing any bony injury as Abbott and I had pointed out in 1953 with regard to the whiplash injury.

Pain in the spine, with or without radiation into an extremity, limitation of neck or back motions, tenderness of the spinal column, or a contusion over the spinal axis, may be present in such injuries.

The measures usually required are rest on a firm bed, application of warmth to the injured part, administration of analgesics and sedatives, and occasionally intermittent light traction or a spinal support. General nervous reactions may accompany these minor spinal injuries. Nervous symptoms can usually be avoided by discussion of the condition with responsible relatives or the patient (in the case of an older child), prompt treatment, frequent reassurance, and early ambulation.

Subluxation

Very severe sprains of the cervical or lumbar spine may permit abnormal motion or alignment of the vertebral column. The symptoms of local pain and tenderness, limitation of motion and muscle spasm are usually very intense and persistent. Both objective signs and subjective symptoms of nerve root irritation may be present. Nerve root irritation or radiculitis is usually manifested by intermittent pain radiating from the site of irritation in accordance with a segmental pattern. This discomfort is commonly accompanied by muscle spasm and is aggravated by mechanical movements of the spinal column or indirect influences on the sensitive nerve roots afforded by coughing, sneezing or straining. Deep tendon reflexes of an affected extremity may be increased when the nerve root has become hyperirritable or decreased when edema and vascular congestion interferes with the normal transmission of the nerve. Altered sensation and hidrosis may occur and these changes follow a segmental dermatome pattern.

In order to classify a spinal injury as a subluxation, some degree of abnormal mobility or alignment of the vertebral bodies must be demonstrated by plain lateral or flexion and extension lateral views on roentgen examination.

Treatment of this condition may be identical with methods employed in case of a simple sprain, but usually the symptoms are more tenacious, and thus the treatment program needs to be more elaborate and prolonged. Light traction is usually necessary until the symptoms of pain and muscle spasm have substantially subsided. Five to ten days of intermittent traction are usually sufficient. A cervical collar or a lumbosacral belt should be obtained as soon as possible so that the patient can be ambulated when not in traction. The neck or back support should be worn intermittently for

about three to six weeks. The interests of the patient are best served physically and psychologically by discontinuing traction, immobilization and spinal supports as soon as possible.

Subluxation of Atlanto-axial Joint

Subluxation of the atlanto-axial joint may occur as a result of trauma or as Sullivan reported a complication of inflammatory conditions affecting the nasopharynx or neck. An intractable torticollis may suggest the presence of this condition. An anteroposterior view of the upper cervical spine taken through the open mouth usually shows a decreased joint space on the affected side and a lateral displacement of the odontoid process toward the affected side. Occasionally, an anterior displacement of the body of the atlas may be seen on the lateral roentgenogram of the upper cervical spine.

The treatment of choice is traction of the head and neck with the cervical spine aligned in a position of slight hyperextension. Either a head halter or skull tongs may be used. After obtaining clinical and roentgenographic evidence of reduction of the subluxation, the traction force may be reduced and the patient ambulated with the neck immobilized by means of a cervical collar. The duration of traction and collar immobilization usually extends from two to six weeks. Laminectomy and decompression of the upper cervical spinal cord may be required in rare instances of intractable subluxation associated with abnormal neurologic signs. A cervical occipital fusion has occasionlly been performed in cases of instability of the atlanto-axial joint but this procedure is rarely necessary.

Fracture of Vertebral Appendages

A simple fracture of a spinous or transverse process or an undisplaced fracture of a lamina usually produces temporary localized pain, tenderness and muscle spasm. No special therapy is necessary other than rest on a firm bed, application of warmth to injured part, relief of pain and early ambulation. A cervical collar, body brace or cast, or lumbosacral belt may be necessary for a short period of time (three to six weeks) when painful symptoms persist.

Fracture of Vertebral Body

Violent flexion of the spinal column may produce a compression fracture of the vertebral body without dislocation. The resulting angulation of the spine may become a permanent deformity unless effective reduction is accomplished promptly. The object of treatment in this condition is to maintain the vertebral column in normal alignment until bony and soft tissue healing has commenced and painful symptoms have subsided. Serial roentgenography is the best method of determining the success of the reduction program and the progress of bony healing.

In compression fractures of the cervical spine, reduction is best accomplished by head and neck traction. The halter method of traction should be commenced as soon as the neck injury is suspected. After the patient recovers from shock, direct skull traction should be substituted for the halter method. Traction is usually required for at least two weeks. A cervical collar or brace may then be worn for an additional period of about four weeks.

Thoracic or lumbar compression fractures are best treated by hyperextension of the spinal column at site of injury. During the first week, a hyperextension cast may be applied and then split into anterior and posterior halves. This measure will enable the patient to be turned without manipulating the injury site and facilitates the nursing care of all parts of the trunk. After about four weeks of immobilization in a position of hyperextension, the patient may be ambulated with an appropriate body brace or lumbosacral brace. Traction is of no value in thoracic injuries. On the other hand, bilateral leg traction may be useful in lumbar fractures for acute or persistent symptoms of pain and muscle spasm.

Intervertebral Disc Protrusion

Protrusion of an intervertebral disc may occur in association with any type of spinal injury.* In general, a disc protrusion is suspected in those cases with spinal radiculitis in which the symptoms and signs are unusually severe and persistent and either partly or wholly disabling. Radicular pain, muscle spasm, alteration of normal vertebral posture, limitation of spinal movement, tenderness of the spinous processes in the affected area, abnormal sensation, muscle weakness and reflex changes are the most common findings in this condition. In cases in which laminectomy is not otherwise indicated, the symptoms of an acute disc protrusion should be treated conservatively. If there are signs of compression of the spinal cord, cauda equina or nerve roots, or if the symptoms of the protrusion are intractable and disabling, myelography followed by laminectomy and removal of the protrusion are indicated. If a laminectomy is performed for the purpose of relieving spinal cord or cauda equina compression caused by displaced bone, the operator should in all instances search for a concomitant disc protrusion at the site of the injury.

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* Editor's Note. A herniated intervertebral disc is most often the result of a degenerative process with symptoms being mild and gradual in onset and with remissions. Progression of symptoms may occur over a period of years However, minor trauma, as lifting, twisting, etc., may precipitate symptoms due to an already degenerated disc. When trauma, such as a fracture dislocation occurs, the posterior annulus tears and the normal disc ruptures, severely compressing nerve roots or spinal cord. The resulting neurological deficit is usually immediate and severe In such instance, trauma is the true etiology of the protruded disc, while in the former instance trauma is only the precipitating force and the basic disease is a degenerative one —I J J

Fracture-dislocation

The most serious spinal injury occurring in infancy and childhood is a fracture-dislocation of the vertebral column. It is imperative to exercise every precaution in such injuries to prevent further aggravation of the injury in handling the patient for purposes of transportation, investigation or treatment. The fracture-dislocation should be reduced as completely as possible, even in cases of complete and irreparable destruction of the spinal cord at the site of the injury.

In general, the same measures described for vertebral fractures are used to align the spinal column in the case of fracture-dislocation. Heavy direct skull traction is used to reduce a fracture-dislocation of the cervical spine, and then the weight is reduced to provide light maintenance traction. A position of hyperextension is usually employed to reduce thoracic and lumbar fracture-dislocation. In lumbar fracture-dislocations, bilateral leg traction may be necessary to relieve symptoms of pain and muscle spasm in addition to hyperextension of the lumbar spine. A spinal fusion is indicated in rare instances of thoracic fracture-dislocation in order to stabilize the site of injury.

Indications for Conservative Management

A conservative program is indicated in the case of most fracture-dislocations even in the presence of profound neurologic deficits below the level of the injury. The following circumstances are definite indications for conservative management:

1. Absence of neurologic deficits.
2. Steady improvement of neurologic signs and symptoms.
3. Unsustained improvement of neurologic signs without evidence of spinal compression by roentgenography or abnormal spinal fluid dynamics.
4. Complete motor and sensory paralysis known to be present since the occurrence of accident.
5. Profound deformity of the spine on roentgen examination which is incompatible with functional survival of the spinal cord at site of injury.
6. When the patient is moribund from intractable shock or overwhelming multiple injuries.

Indications for Laminectomy

Whenever there is indisputable evidence of compression of the spinal cord or cauda equina, a laminectomy is indicated. Some of the following conditions constitute such evidence:

1. Neurologic deficits below site of injury which are increasing in variety or intensity.

2. Roentgenographic evidence of marked narrowing of the spinal canal.
3. Spinal subarachnoid block whether partial or complete.
4. Partial recovery of spinal function below injured area combined with roentgenographic evidence of bony displacement or partial subarachnoid spinal block.
5. Intractable nerve root pain originating near site of injury.
6. Evidence of compression of nerve root or cauda equina by myelography.

Suggestions Regarding Laminectomy and Spinal Decompression

Whenever a laminectomy is indicated, it should be performed as soon as the patient can tolerate the administration of a general anesthetic. The service of an anesthesiologist should be obtained if such a specialist is available. It is important to employ an endotracheal tube. A sample of blood from the patient should be typed and cross-matched with blood from a compatible donor before commencing the anesthesia. It is recommended that a pediatrician examine the patient before operation and follow the patient daily in the postoperative period.

In most instances the operation is performed with the child lying in a prone position and both shoulders supported by bath blanket rolls. The chief indication for operating with the patient on one side arises in case of a chest injury or complicating cardiac or respiratory condition. A special neurosurgical head support is necessary for performing a laminectomy in the cervical or upper thoracic region.

The incision is made in the midline of the spine and centered over the site of injury. The location of the injury may be obvious due to deformity of the spine or ecchymosis of the skin. However it is advisable to view the roentgenograms in the operating room in order to plan accurate placement of the incision. In general the skin incision is relatively longer in children than in adults and should be extended for about three vertebrae above and three vertebrae below the principal site of spinal compression.

The muscles and fascia are separated by sharp dissection and retracted from the spines and laminae on both sides. Unusual care must be exercised to prevent injury of neural elements by instruments or manipulation of bone fragments in case of a fractured lamina, ruptured ligamentum flavum or congenital spina bifida occulta. The spines, laminae and intervening ligaments are removed from at least two vertebrae above and two vertebrae below the fracture-dislocation. It is advisable to preserve the vertebral facets whenever possible.

A search is made for possible protrusion of an intervertebral disc or bone fragment from a vertebral body impinging on the spinal canal. If these conditions are found, they must be carefully removed.

The dura is seldom opened. The chief indication for exposing the cord occurs in instances of suspected hematoma of the spinal cord. If no compression of the spinal cord is found, the dura should be opened and the spinal cord inspected. On rare occasions it has been possible to evacuate an intramedullary hematoma by cautious aspiration or incision of the spinal cord.

The wound is closed with interrupted sutures inserted in each soft tissue layer. Drains will be indicated in rare situations where bleeding has been controlled with extreme difficulty and postoperative hemorrhage may be a serious threat, but most neurosurgeons never drain such spinal wounds.

Transportation

Pain in the neck or back, tenderness or bruising of the spinal column, deformity of the neck or back, or weakness and loss of sensation in the extremities, may suggest the possibility of spinal injury. The injured infant or child must be rolled or lifted onto a firm stretcher or board by a group of rescuers, taking exceptional precautions to prevent flexion or extension of any portion of the spinal column. The patient must be prevented from turning or bending the neck or trunk while en route to the hospital. When a neck injury is suspected, the patient is placed in a supine position and a standard head halter should be placed about the head and light traction applied to the spreader. The head halter and other appropriate tackle belong among the equipment items in ambulances and in the physician's bag. The same thoughtful management of patients with a suspected spinal injury is necessary in the emergency room, department of roentgenology, and when handling the patient anywhere in the hospital. Such precautions will effectively prevent additional trauma to spinal structures in the interval between rescue and definitive treatment in the hospital.

Management of Patient With Acute Spinal Injury

Emergency Treatment

On admission to the hospital, the exact condition of the patient must be promptly and accurately established. A search is made for all types of injuries and the degree of shock is estimated. The establishment of unobstructed respirations, control of bleeding and measures to reduce shock take precedence over special examinations and treatment of other conditions. The patient should be kept warm, and oxygen administered by nasal catheter. An unobstructed airway is established by positioning the head, insertion of a pharyngeal or nasal airway, or an intratracheal tube or performing an emergency tracheotomy. Bleeding can usually be controlled by pressure dressings. If the condition of the patient is poor, fluids should be

administered at once by the intravenous route. In the meantime, a specimen of blood from the patient may be typed and cross-matched with the blood of a suitable donor so that a transfusion of whole blood may be administered with the least possible delay. It is possible to carry out this emergency treatment program and still manage the patient in such a manner as to avoid jeopardizing the spinal injury. Restless or irrational patients may require restraints and sedation.

Examination

A general physical and neurologic examination may be carried out as soon as the patient's general condition improves. The temperature, pulse, respirations and blood pressure of the patient should be taken at regular intervals and a record maintained of these and other pertinent observations. The general physical examination should consist of inspection of the entire body, palpation of the spine, chest, pelvis and long bones, auscultation of the heart and lungs, and manual examination of the abdomen. Essential neurologic information may be obtained by observing the state of consciousness and function of the cranial nerves, testing of deep tendon and abdominal reflexes, noting the movement of the extremities in response to commands or painful stimulation, and testing for intactness of pain sensation in all parts of the body. In cases where gross neurologic deficits are present, special attention should be directed to detect any gastrointestinal and bladder distension.

Roentgenography

When the injury is mild or the condition of the patient is stable, the patient should be transported to the department of roentgenology for examination. Although portable roentgenograms are notoriously inadequate in a study of the spine, this may be the only method that is practical in the case of a patient who is in critical condition. An anteroposterior and lateral view of the suspected site of spinal injury are essential. Special views of the spine should usually be delayed until the condition of the patient has improved.

In suspected subluxation of the atlanto-axial joint, an anteroposterior view taken through the open mouth will be useful. A complete cervical roentgenographic study consists of anteroposterior, lateral, right and left oblique, and flexion and extension lateral views. There is usually considerable muscle spasm associated with cervical injuries so that the patient has his shoulders raised cephaled. In such instances, the lower portion of the cervical spine will not be seen in the later roentgen views. Since the lower cervical spine is most frequently involved it is imperative to visualize this area. Therefore, manual traction is applied to the head and counter-traction to both

arms thereby pulling the shoulders down and out of the way allowing the lateral x-ray to include the lower cervical vertebrae in sharp detail. Only anteroposterior and lateral views are useful in case of injury of the thoracic spine. A complete study of the lumbar spine consists of anteroposterior, lateral, and right and left oblique views.

Lumbar Puncture

It is useful to perform a lumbar puncture in the case of all major spinal injuries which involve a fracture of a lamina or vertebral body, fracture-dislocation of a vertebra, or when neurological deficits are present. The patient should be carefully rolled onto one side with the head supported by a pillow, and legs extended. The fourth lumbar interspace is the most practical site for the puncture. An 18 gauge needle should always be used. Once the puncture has been achieved, a water manometer is employed to measure the initial pressure. The appearance of the fluid is noted. After an acute spinal injury, the cerebrospinal fluid may be clear or contain blood. Bloody spinal fluids are more common in cases of severe types of spinal cord injury (contusion or laceration). The jugular compression test (Queckenstedt test) is next performed. An assistant compresses both jugular veins in the lower neck for five seconds while the physician observes the response of the cerebrospinal fluid column in the manometer. A prompt rise during compression and a prompt fall of the column after the jugular compression has been released indicates that no compression of the spinal subarachnoid space is present at the injury site. A very slow rise and fall of the fluid column indicates a partial block. If there is no response of the column during jugular compression, or a slow rise and no fall of the column, a complete block is suspected. In order to verify the presence of a partial or complete subarachnoid block, it is imperative to check the patency of the needle by observing the response of the column to abdominal pressure. Since a different mechanism is involved, a prompt rise and fall of the column will be observed during abdominal pressure as long as the needle is patent. A block of the subarachnoid space may be caused by bony compression, hematoma, or swelling of the spinal cord. The finding of a partial or complete block of the subarachnoid space by manometry represents only one of the factors that enters into the decision as to whether a laminectomy should be performed.

Treatment

A general outline of treatment for specific conditions is contained in the paragraphs describing individual types of spinal injury. The purpose of this section is to present the details of the methods to be followed. It should be

pointed out that all spinal injuries provoke profound anxiety and excitement on the part of the patient, relatives, public authorities and even hospital personnel. It is important for the physician to provoke confidence on the part of all concerned by attending the injured promptly, and effectively treating the injuries.

Spinal Traction

I. *Purpose.* Traction may be applied to the head and neck, pelvis or lower extremities in the case of cervical or lumbar spinal injuries in order to accomplish some or all of the following objectives:

A. *General*

1. Enforce rest
2. Reduce muscle spasm
3. Relieve pain from nerve root and muscle sources
4. Psychological advantage of an active and physical treatment of injured part.

B. *Special*

1. Align and immobilize the vertebral column to facilitate soft tissue and bony healing.
2. Reduce a fracture-dislocation.

II. *Principles:* In order for traction to be effective, the following principles must be considered:

1. The method selected must accomplish the desired general and special objectives.
2. Reasonable comfort for the patient must be assured.
3. In case of serious spinal injuries, the method should allow rapid application with a minimum of shock or manipulation of the injured part.
4. In minor spinal injuries, it should be easily applied and removed to allow ambulation of patient when out of traction or application in home environment.

III. *Methods of Cervical Traction*

1. *Fronto-occipital Halter.* A bandage (gauze, flannel or felt-backed sponge rubber) may be applied snugly around the circumference of the head. Traction straps of the same material may be tied or pinned to the headband and then fastened together to form a traction loop to which a rope and weight is attached as described by Love over twenty years ago. This method is useful in first aid practice but has the disadvantage of causing uncomfortable pressure against the forehead and temples, and headache.
2. *Occipito-mental Halter* (Figures 1 and 2). This type of halter



FIGURE 1. (Left) Occipito-mental halter traction in use. Note the angle of pull (about forty-five degrees). Four lbs. of weight usually provides effective maintenance traction in children.

FIGURE 2. *(Right)* Improvised occipito-mental halter constructed of felt backed sponge rubber (with slit in center), wooden spreader, rope and metal ring. This apparatus often is more comfortable than commercial models.



FIGURE 3. Photograph of manikin illustrating method of direct skull traction, applicable in severe cervical spine injuries in infants. Loops of wire passed through burr openings (placed opposite mastoid process) may be insulated with polyethylene tubing to prevent eroding the soft cranium.

with metal spreader is commonly used for temporary or intermittent cervical traction. When carefully secured to the chin and occiput and connecting straps have been fastened evenly, this method is very effective. The chief disadvantage has been the discomfort caused by pressure against the chin, mandible and tempero-mandibular joint. Many models have been improperly constructed so that care should be exercised to prescribe the most comfortable and durable type available.

3. *Direct Skull Traction in Infants.* Traction may be applied directly to the skull by passing a loop of stainless steel wire between two burr openings in the skull. A pair of steel wire loops may be placed at symmetrical locations on each side of the midline of the skull and then attached to a spreader. This method has been used to provide prolonged or heavy traction in infants and children under six years of age. This is the method of choice in all children in whom the thinness of the skull prevents the use of head tongs. It is both a painless and effective method. The chief technical problems include preventing the wires from cutting through the soft bone and se-

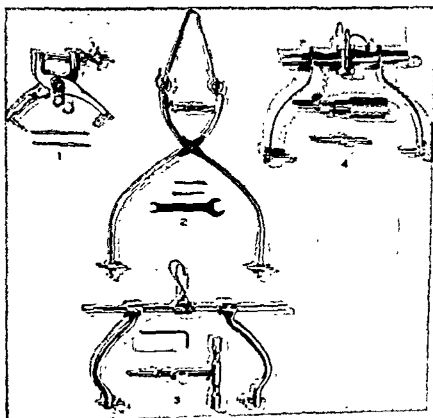


FIGURE 4. Four types of tongs satisfactory for direct skull traction in older children suffering from severe cervical spine injuries. (1) Crutchfield type (2) Barton type (3) Blackburn type (4) Vinke type.

lecting wires that will not break under ordinary torsion or stretch. These mishaps can be prevented by insulating the wire with polyethylene tubing where it passes between the burr openings, using moderate traction force and employing heavy stainless steel wire (Figure 3).

4. *Direct Skull Traction in Children.* The Barton skull traction apparatus is the most suitable instrument for accomplishing prolonged cervical traction in children over six years of age. In children of teen age, any of the other standard tongs (Crutchfield, Blackburn or Vinke types) may be employed providing the thickness of the skull permits their use. The skilled neurosurgeon should be familiar with the application of all types of tongs (Figures 4 and 5).



FIGURE 5. Roentgenogram of skull showing application of Barton tongs in case of a six year old child suffering from atlanto-axial dislocation as result of being struck by an automobile. Most prominent neurological finding was bilateral hypoglossal nerve paralysis. The patient made a complete recovery. (*Roentgenogram courtesy* Dr. Robert M. Jaeger)

5. *Other Methods.* Two other methods should be mentioned since each could be used as an improvisation in case of disaster. One employs a pair of barbless fishhooks placed beneath the zygomatic processes. The other method consists of a pin driven through the ligamentum nuchae.

IV. *Lumbar Traction*

A. *Pelvic Traction.* A lumbosacral belt with attached traction straps may be employed to provide lumbar traction. This method has the advantage of being comfortable and easily applied or removed. In clinical practice, it has been less effective than bilateral leg traction.

B. *Bilateral Leg Traction.* Most of the older methods employed ad-

hesive materials in order to apply traction to the legs. The best method now available is Gershman's, which consists of binding strips of felt-backed sponge rubber to each leg with elastic bandages. Each leg should have a separate spreader and weight system.



FIGURE 6. Practical method of applying bilateral leg traction consisting of felt-backed sponge rubber secured by elastic bandages (without applying adhesive substances to the skin). Note the simple type of pulley suspension applicable for hospital or home use, when either cervical or lumbar traction may be prescribed.

tem. Four to 6 lbs. pull on each leg is usually adequate (Figure 6).

V. *Traction Control*

1. The position of the patient in bed should be controlled by elevating the head of the bed in case of cervical traction, or elevating foot of bed in pelvic or lumbar traction.
2. The direction of pull should be adjusted to maintain the cervical or lumbar spine in a neutral position. Portable roentgenography may be employed to check the vertebral alignment and direction of pull.
3. The amount of weight applied to the head and neck, pelvis, or legs, should be the minimum necessary to achieve the objectives of the method and within the comfort tolerance of the patient. Light traction (7 lbs. or less) is usually adequate in minor spinal injuries, high cervical fracture-dislocations, lumbar injuries and in cases of injured infants and young children. Heavy traction (over 7 lbs.) has been required for major spinal injuries, low cervical fracture-dislocations, and spinal injuries

in older children. The minimum maintenance traction weight is 4 lbs., and the maximum weight required to reduce the most stubborn fracture-dislocation is 35 lbs.

4. In general, light traction (4 to 7 lbs.) should be initially applied. The weight may be progressively increased until the desired result has been obtained, and then the weight may be reduced to the minimum amount of weight required to maintain the desired objective. Portable roentgenography must be employed to measure progress in traction reducing a fracture or fracture-dislocation. Once traction is applied, one form of definitive treatment has been started. It is not necessary to accomplish all objectives of traction immediately. Best results are obtained by reducing a severe fracture dislocation in a few hours (not minutes).

Hyperextension of Spine

Hyperextension of the spine has been the treatment of choice in uncomplicated thoracic and high lumbar spinal injuries. This position can be achieved at once by placing a folded blanket beneath the patient or mattress opposite the injured part (Figure 7). Prolonged hyperextension is usually required to allow bony healing and is desirable in order to allow early ambulation. An orthopedist or brace maker should be enlisted in the treatment program to provide a suitable body cast or brace.

The duration of hyperextension should be controlled by serial roentgeno-

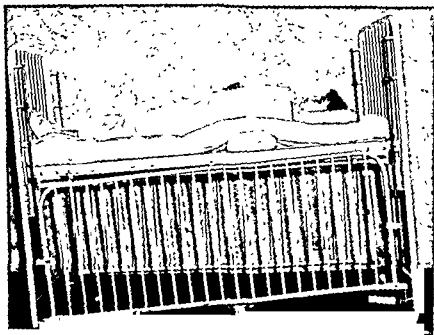


FIGURE 7. Satisfactory method of maintaining the spine in hyperextension. This position useful in vertebral body fractures and fracture-dislocations of thoracic or lumbar spine.

grams of the spinal injury site. Once evidence of bony healing is obtained, the cast or brace may be discarded.

Spinal Supports

Some type of spinal support may be required in order to allow early ambulation or insure comfort during convalescence following severe spinal injuries. In most instances, only partial immobilization of the spine is intended in order to allow some movement of the spinal musculature, and to prevent extensive atrophy of muscles. In addition, excessive immobilization or prolonged use of spinal supports is more likely to result in unfavorable psychoneurotic reactions.

A cervical collar, constructed of corrugated paper or plastic material with appropriate fitting, padding, covering and fastening, is suitable for use in most spinal injuries for partial and temporary support of the cervical spine. Improvised collars allow some movement of neck muscles and are likely to wear out before the patient develops extreme muscular atrophy (Figure 8).

When there has been a severe bony injury, such as fracture or fracture-



FIGURE 8 Improvised cervical collar constructed of corrugated paper, sheet wadding, stockinet and adhesive tape Adequate temporary neck support obtained by this method, and collar wears out before atrophy of muscles occurs.

dislocation of the cervical spine, or in rare instances when reduction of the fracture has been difficult or intractable, a commercial brace support of the head and neck should be employed.

In thoracic and lumbar vertebral fractures or fracture-dislocations, a body brace may be necessary in order to permit ambulation early in the convalescent period.

Rest on Firm Bed

It is important to place a patient with a spinal injury on a firm bed. Most hospital beds in the pediatric department have a strong spring base and firm felt mattress; *however, in case of a sagging bed, it will be necessary to* use a bedboard of half-inch plywood between the mattress and springs.

Application of Warmth

Warmth applied to a spinal injury site is useful as a symptomatic measure for the relief of pain and muscle spasm and in order to increase the efficiency of affected muscles. The use of dry warmth in the form of an infra-red appliance is recommended. The most important consideration is the use of mild warmth rather than intense heat in order to obtain the maximum symptomatic relief. It is best to apply warmth twice a day for a period of 2 to 4 weeks.

Analgesics and Sedatives

Most patients who have suffered a spinal injury require analgesics and sedatives after they have recovered from the initial shock of the injury. Aspirin and phenobarbital can usually be relied upon to make the patient more comfortable and relaxed.

Early Ambulation

The patient should be ambulated at the earliest possible moment. In mild injuries, ambulation can begin as soon as the acute symptoms have substantially subsided. In more severe injuries, ambulation should be delayed until the bony and soft tissue healing has effectively commenced and the skeleton is stabilized. If a patient is encouraged to increase his activities rapidly, muscular atrophy may be minimized and the morale of the patient is enhanced. Many treatment programs are too inflexible and unnecessarily prolonged. *The patient is benefited by individualizing the* treatment program and adapting a courageous attitude of reestablishing the individual's normal activities as completely as possible with the least possible delay.

Rehabilitation

In case of a mild spinal injury, the resumption of normal activities may be all the rehabilitation that is required. In more severe injuries, the services

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Rehabilitation

In case of a mild spinal injury, the resumption of normal activities may be all the rehabilitation that is required. In more severe injuries, the services

of a physiatrist may be advantageously employed to direct the program of rehabilitation. As soon as the patient has recovered from the shock caused by spinal injury, physical therapy may be commenced. It is often possible to begin active or passive movements and massage of affected muscles as early as the day following the occurrence of the injury. Physical therapeutic measures should be considered part of the patient's program even in the acute phase of the problem.

Miscellaneous Measures

In addition to the management of shock, when the injury is acute, and the reduction of the fracture or fracture-dislocation, Monro described many technical details that are important in the management of a patient suffering from paraplegia or quadraplegia. Such patients should be maintained on a firm bed with dry, clean and wrinkle-free bedding. The patient's position must be changed every two hours from back to side to other side. Bedding and clothing should be clean and dry and changed frequently. The skin should be massaged several times during a twenty-four hour period. The hips, back, sacrum and heels are inspected frequently for signs of redness, excoriation or ulceration. Whenever abnormal changes in the skin occur, every effort should be made to keep weight off of those sites.

An indwelling catheter will be required. The simplest procedure is to employ either intermittent or continuous gravity drainage and to irrigate the bladder with sterile water twice daily. However, the use of a tidal drainage apparatus is preferred when this is available and personnel are familiar with the method. Each week, a urinalysis and urine culture should be ordered and the catheter should be changed. An appropriate antibiotic may be administered as prophylaxis against urinary infection. The services of a urologist should be enlisted in the program when such a specialist is available.

It is useful to maintain a record of fluid intake and output. When the injury is acute, it is advisable to administer fluids by the intravenous route in order to avoid distension of the gastrointestinal tract which is prone to occur with mouth feeding. Distension of the gastrointestinal tract can often be avoided by prohibiting all fluids and foods by mouth for the first few days. If distension develops, the stomach must be kept emptied by gastric suction and the large intestine decompressed by the intermittent use of a rectal tube.

After peristalsis has returned, the patient may be fed by mouth. It is usual for loss of weight to occur, which is due in part to loss of appetite, discomfort and inactivity. For this reason, high vitamin and caloric intake is advised. The services of a dietician are usually necessary in order to maintain the best state of nutrition in the patient.

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CHAPTER XVIII

Trauma—Peripheral Nerves

Robert S. Knighton

With the exception of injuries incident to a traumatic birth, peripheral nerve injuries in childhood differ in no essential respect from those incurred in adult life. In a review of sixty-five cases of peripheral nerve injury in childhood occurring between 1942 and 1955, a variety of etiological agents was elicited. These included lacerations by falling upon broken glass, tin cans, or from automobile accidents; fractures from falling on the outstretched hand, or from automobile accidents; stretch injuries from falls upon the shoulder or abnormal pulling of the arm; accidental gunshot wound, and other miscellaneous accidents. Severance of the nerves in the forearm by falls on broken glass occurred with a surprisingly high frequency in children between the ages of eight and twelve years, and was the most frequent statistical cause of peripheral nerve injury.

In general, the technique of treatment of each individual nerve lesion in children follows essentially the same principles as it does in adults. While no detailed comparison could be made between the functional result after nerve repair in children and adults, the impression upon reviewing this series of nerve injuries in children was that the return of function following a good nerve repair, particularly of the median or ulnar nerve at the wrist, was greater than with a similar case in adults. In several instances, examination many years after the original injury and nerve suture elicited no trace of nerve injury. On the other hand, when regeneration was poor, particularly with injuries of the brachial plexus or sciatic nerve, the factor of retarded growth of the extremity was added to the deficit already present from the nerve injury and thereby made the deficit greater than with a comparable injury in adult life.

Injuries at Birth

Delayed or precipitous labor, particularly with abnormal presentation, or the use of forceps, is responsible for the majority of injuries to the peripheral nervous system occurring at the time of birth. The facial nerve and brachial plexus are the structures most commonly traumatized.

Injury of the Facial Nerve

This is a relatively frequent complication of delivery. In most instances it is caused by compression of the branches of the facial nerve by forceps, but may also occur from prolonged pressure of the face against the pelvis in difficult labor, or from the rapid passage of the child through the birth canal during precipitous labor. The interruption of the nerve in most instances is physiological and usually not complete. The period of recovery which usually is complete, varies from a few days to a few months. The condition is readily recognized by the inability of the child to close the eye and the asymmetry of the mouth. Nursing usually is not a problem. Treatment is usually not necessary. If there is no recovery within a year, a hypoglossal facial or accessory facial repair is indicated.

Injury to the Brachial Plexus

This type of injury is produced by stretching of the trunks of the brachial plexus, or of the nerve roots forming it. It may occur either during breech delivery, if traction is applied to the shoulder, or in cephalic presentation if the head is drawn too forcibly away from the shoulder. Both of these maneuvers result in the upper arm, or Erb's type of palsy. The upper trunk of the brachial plexus, or the fifth and sixth cervical nerve roots forming it, are the structures most commonly involved, although the lesion may be more extensive, depending upon the degree of force exerted.

Maneuvers which hyperextend and cause traction on the arm in the hyperextended position are thought to be responsible for the rare lower arm and hand or Klumpke type of palsy. The involvement in this type of palsy is primarily in the lower cervical and uppermost thoracic nerve roots, or the trunks formed from them.

The damage to the peripheral nerves in either of these injuries may be in the spinal canal and consist of root avulsion from the spinal cord, or it may be distal to the spinal canal, either in the roots or trunks forming the brachial plexus, or it may be a combination of these. The recognition of either of these lesions is not difficult because of the typical posture assumed by the extremity. Early in both types of injury the arm appears to be flaccid without voluntary motion on the part of the child. The reflexes are usually absent and often response to painful stimulation is likewise absent. If the root avulsion is higher than the fifth cervical nerve root, paralysis of the diaphragm on the affected side may be present resulting in some difficulty with respiration. Later the arm and hand assume a posture characteristic of the type of palsy present. This posture is caused by the unopposed action of the muscles unaffected by the injury. In Erb's palsy the arm is adducted and internally rotated. The forearm is extended and pronated, and wrist drop is present. In the Klumpke type of palsy, which

is extremely rare, the musculature supplied by the seventh, eighth and first *thoracic nerve roots is involved* producing paralysis primarily in the musculature of the hand. This produces a claw-like appearance of the hand. In addition to this, there are also evidences of trophic disturbance and sometimes a Horner syndrome due to involvement of the sympathetic innervation to the cervical plexus.

The treatment of cervical root and brachial plexus injuries, as described above, is non-surgical and consists essentially of proper splinting early to prevent overstretching of the paralyzed muscles combined with proper physical rehabilitative procedures in the nature of massage and passive motion. Later, as some voluntary function returns, and the child is old enough to cooperate, the addition of muscle re-education is added to the treatment. The degree of functional return is predetermined by the nature of the injury, and fortunately, in most instances of the upper arm type, the outlook is reasonably favorable. Since the lower arm type is caused by a more severe injury, and is also more likely to be the result of root avulsion than a stretch injury to the brachial plexus, the outlook is less favorable. Reconstructive surgery, in the nature of tendon transplantation, may be of some value in the later treatment of these injuries.

Injuries in Childhood

Injury to the cranial and spinal nerves may occur at any age after birth, but, as already mentioned, is most frequently encountered near the end of the first decade of life.

Injury to the Cranial Nerves

For the most part injuries to the cranial nerves are produced by head trauma. Those most commonly involved are the olfactory, due to injuries producing contusion of the orbital surfaces of the frontal lobe; the optic, due to fracture extending through the optic foramen; the vestibular, auditory, and facial nerves from fracture of the petrous bone. Injury to the facial nerve and its peripheral branches may also occur with lacerations about the face and is important from a surgical standpoint, as prompt suture offers the best opportunity for a successful return of function. Hypoglossal facial anastomosis is indicated if facial nerve function fails to return after fracture through the petrous bone.

Injuries to Spinal Nerves

As mentioned previously, injuries to spinal nerves may be caused by a variety of traumatic agents. The recognition of nerve involvement is not difficult when anticipated. Appraisal of the site of injury, the agent responsible for it, and an evaluation of *the muscle and sensory function supplied* by the nerves in the area, permit an accurate diagnosis. Since many of these

lesions, particularly at the wrist, involve tendons and arteries, as well as nerves, the exact nature of the injury may not be apparent until surgical exploration of the wound is made. However, a definite clue that nerve involvement is present can always be obtained beforehand so that this important part of the restoration of structures in the injured extremity is not overlooked.

In our review of nerve injuries the median and ulnar nerves were by far the most frequently involved, and in practically all instances, there was associated tendon, and often major blood vessel injury, as well. Most of these injuries occurred at, or near, the wrist and were, for the most part, caused by broken glass and sharp metal, such as a tin can lid or knife. Nerves in the proximal portion of the upper extremities, and in the lower extremities, were most likely to be injured by closed trauma, such as fracture or contusion, compression by casts or stretching.

In testing the function of muscles, many factors must be taken into account. The more important of these perhaps are: (1) the ability of the patient to cooperate; (2) the presence of severed tendons, as well as nerves; (3) the influence of gravity on testing of muscle function, and (4) the presence of supplementary movements which may be due to anomalous nerve supply or accessory tendinous insertions, or by rebound of paralyzed muscles by sudden release of strongly contracting antagonist muscles.

Brachial Plexus

Injuries to the brachial plexus in children are most commonly a result of stretch or avulsion accidents, often with associated fracture of the clavicle, or fracture dislocation about the shoulder. A minor, though sometimes alarming, type of stretch injury is occasionally seen in infants as the result of rough play during which the arm is abnormally stretched, particularly while in extension, or when the child is forcibly lifted from the floor by his arm. For several days, or even weeks, the child refuses to move the arm and the reflexes may be lost. Fortunately, these injuries are usually not permanent and ultimate complete recovery is the rule. The musculature most commonly involved is that supplied by the upper roots and trunk of the brachial plexus. The degree and site of injury can usually be assessed with fair accuracy by examination of the muscle function, reflexes and cutaneous sensation. Lumbar puncture may also aid in the evaluation of the injury, as bloody spinal fluid suggests that the lesion is due, at least in part, to avulsion of the nerve roots from the spinal cord.

Axillary Nerve

This nerve is occasionally injured following fracture dislocation of the shoulder, or fracture of the upper end of the humerus. It innervates the deltoid muscle, which is the principal abductor of the arm to the hori-

zontal position. Injury to this nerve is detected by inability to perform this function and by a zone of hypesthesia of the skin overlying the lower two-thirds of the posterior aspect of the deltoid muscle.

Musculo-cutaneous Nerve

This nerve is rarely injured in children because it is well protected by the biceps muscle which it supplies. Loss of function of this nerve is detected by inability to flex the forearm and by a zone of hypesthesia on the radial surface of the forearm.

Radial Nerve

Injury to the radial nerve most commonly occurs as a result of fracture near the junction of the middle and lower third of the humerus. It may also be injured by fracture dislocation about the elbow joint, particularly dislocation of the head of the radius, and occasionally results from deep lacerations about the upper forearm and arm. While this nerve is less frequently involved by injury than the median and ulnar nerves, it is the nerve most likely to be injured by fracture. Injury to the radial nerve at various levels in the arm and forearm can be detected by examination of the muscles supplied by this nerve. A detailed list of the muscles innervated by this and other peripheral nerves and their primary function may be obtained by referring to Table I. If the injury to the radial nerve is high in the axilla, all of the muscles supplied by it, including the triceps, are paralyzed. If the injury is near the mid humerus, triceps function is usually spared and the most proximal muscle involved is the brachioradialis. In the lower third of the humerus, brachioradialis and triceps function are preserved but dorsiflexion of the hand at the wrist and the fingers at the metacarpophalangeal joints is lost. If the injury is in the proximal portion of the forearm, some of the deep branches might be spared and wrist drop not complete. In all complete injuries of the radial nerve, regardless of level, a zone of anesthesia is usually apparent on the dorsum of the hand between the first and second metacarpals.

Median Nerve

This nerve is most commonly injured by laceration of the forearm at the wrist. Occasionally, however, it is injured by fracture near the lower end of the humerus, or dislocation of the elbow. When severed at the wrist there is usually associated laceration of the palmaris longus, flexor digitorum sublimis, and flexor carpi radialis tendons. In deeper lacerations the tendon of the flexor pollicis longus and the radial artery may also be injured. If the nerve is severed or injured in the arm proximal to the origin of the muscular branches, the lesion is a complete one with paralysis of all muscles

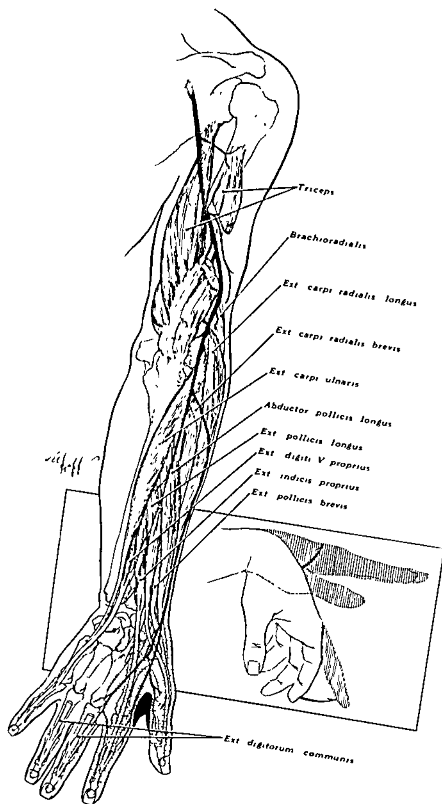


FIGURE 1. Radial nerve showing principal muscles innervated and major tests of function.

supplied by the nerve, as well as complete sensory loss in its cutaneous distribution. A simple test for recognition of median nerve palsy in the arm is to have the patient attempt to flex the distal phalanx when the remainder of the finger is held rigid by the examiner. Inability to do this implies loss of innervation to the flexor digitorum profundus muscle. In addition to this function, opposition of the thumb and fingers is not possible. Sensation to the tip of the index finger, which is the absolute zone of the median cutaneous distribution, is likewise lost. When the lesion is at the wrist, the findings are the same except for flexion of the distal phalanx, which is preserved.

Ulnar Nerve

Like the median nerve, this nerve is most commonly injured by laceration of the forearm near the wrist. It may also be injured by fractures involving

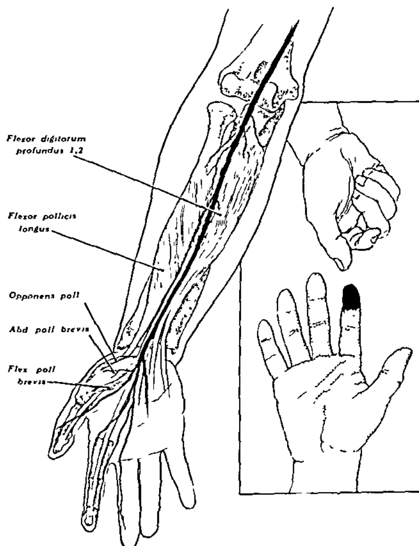


FIGURE 2 Median nerve showing principal muscles innervated, typical posturing of hand and area of sensory loss when nerve is severed

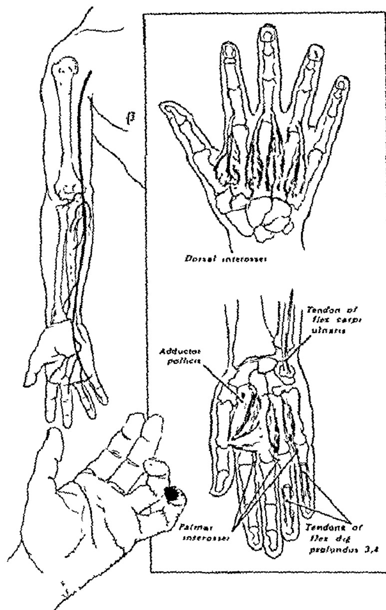


FIGURE 3. Ulnar nerve showing principal muscles innervated, typical posturing of hand and area of sensory loss when nerve is severed.

the lower end of the humerus and particularly the medial epicondyle. Injury to the ulnar nerve in the arm is recognized by loss of function of all of the muscles supplied by this nerve, including the flexor carpi ulnaris and ulnar side of the flexor digitorum profundus. High lesions are recognizable by inability to flex the distal phalanx of the little finger when the remainder of it is held rigid by the examiner and impairment in ulnar deviation of the hand. In lesions at the wrist these two functions are spared but the remainder of the musculature in the hand supplied by the ulnar nerve is paralyzed. This is recognized by the typical appearance of the ring and small fingers which are flexed and separated from one another, the little finger being abducted. When the hand is placed on a flat surface

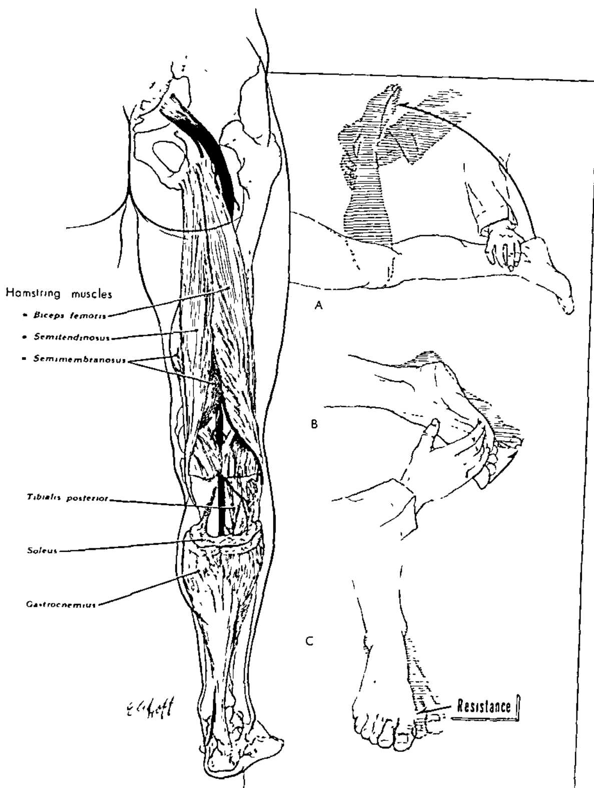


FIGURE 4 Sciatic and posterior tibial nerves showing principal muscles innervated and tests of function (A) Sciatic nerve. (B) and (C) Posterior tibial component.

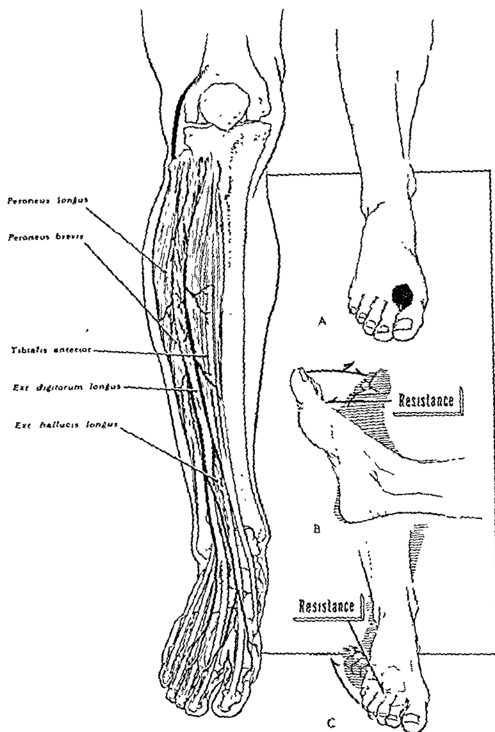


FIGURE 5 Peroneal nerve showing principal muscles innervated by its superficial and deep branches (A) Area of sensory loss following severance of the common peroneal nerve. (B) Tests of function for deep branches, and (C) Superficial branches.

Nerve	*Principal Muscles*	*Principal Function*
Axillary	Deltoid (C5,6)	Abduction of the arm to horizontal
Musculocutaneous	Biceps brachii (C5,6)	Flexion and supination of forearm
Radial		
Upper third of arm	Triceps (C6,7)	Extension of forearm
Lower third of arm	Brachioradialis (C5,6)	Flexion and supination of forearm
At elbow	Extensor carpi radialis longus (C6,7)	Extension of wrist
	Extensor carpi ulnaris (C7)	Extension of wrist
	Extensor digitorum communis	Extension of proximal phalanges and wrist
	Extensor pollicis longus	Extension of terminal phalanx of thumb
	Extensor pollicis brevis	Extension of proximal phalanx of thumb
	Abductor pollicis longus	Abduction of thumb
Median		
At elbow	Pronator teres (C6)	Pronation of forearm
	Flexor carpi radialis (C6)	Flexion and abduction of wrist
	Flexor digitorum sublimus (C7,8,T1)	Flexion of wrist, middle and proximal phalanges
	Palmaris longus (C7,8,T1)	Flexion of wrist
	Flexor pollicis longus (C8)	Flexion of distal and proximal phalanges of thumb
	Flexor digitorum profundis 1,2 (C7,8)	Flexion distal phalanx of index and middle fingers

Nerve	Muscles	Roots	Principal function
At wrist	Abductor pollicis brevis Flexor pollicis brevis (sup. head) Opponens pollicis	(C6,7)	Rotate and flex proximal phalanx and metacarpal of thumb into position of opposition to other fingers
Ulnar At elbow	Flexor digitorum profundis 3,4 Flexor carpi ulnaris	(C7,8,T1)	Flexion of distal phalanx of ring and little fingers Flexion and adduction of wrist
At wrist	Adductor pollicis Dorsal interossei Volar interossei	(C8)	Adduction of thumb to hand Abduction of metacarpals Adduction of metacarpals
Sciatic	Semimembranosus Semitendinosus Biceps femoris	(S1,2)	Flexion of leg upon thigh
Tibial	Flexor digitorum longus and brevis Flexor hallucis longus Gastrocnemius Soleus	(S1,2)	Flexion of foot and toes
	Abductor hallucis Tibialis posterior		Inversion of foot
Peroneal	Extensor digitorum longus Extensor hallucis longus Tibialis anticus	(L4,5)	Dorsiflexion of foot and toes
	Peroneus longus Peroneus brevis	(L4,5)	Eversion and dorsiflexion of foot

FIGURE 6. The peripheral nerves with the specific muscles innervated and principal function.

there is inability to separate or draw the fingers together due to a paralysis of the interossei and sensation on the little finger and ulnar half of the fourth finger is lost.

Sciatic Nerve

Injury to the sciatic nerve is uncommon but occasionally occurs with deep lacerations about the buttocks or posterior thigh and fractures of the femur or dislocation of the hip. Complete high sciatic section produces total paralysis of the foot and loss of flexion of the leg on the thigh. This, of course, is due to the loss of nerve supply to the hamstring muscles and extensors and flexors of the foot and toes.

Tibial Nerve

Injury to the tibial nerve results in loss of flexion of the foot and toes. This injury is uncommon in children and results from a chance injury to this nerve, usually in the popliteal space.

Peroneal Nerve

Injury of the common peroneal nerve, or of the uppermost portion of the deep peroneal nerve, results in a complete foot drop. This injury may be caused by adduction injuries of the knee with fracture of the tibia and fibula. It also occurs occasionally following application of casts and following contusions or lacerations to the nerve in its superficial course across the fibula. If the lesion is distal to the division of the common peroneal nerve, the deep or superficial branches might be injured independently with concomitant loss of the function of the muscles supplied by the involved branch.

Ancillary Tests for Peripheral Nerve Impairment

In addition to the evaluation of motor and sensory function, which is usually all that is necessary for the diagnosis of a completely severed nerve in the acute stage, mention should also be made of ancillary measures which are particularly helpful when the patient is seen for the first time at an interval following nerve injury.

The most important of these is perhaps the response of each muscle to electrical stimulation of its nerve. This can be carried out by applying the stimulating electrode over the point where a nerve trunk passes close beneath the skin, or, if necessary, can be performed with the use of needle electrodes introduced directly into the nerve trunk through the skin. If surgery is decided upon, stimulation, of course, can be carried out under direct observation at the time of surgery. This test is extremely helpful in evaluating whether the nerve lesion is complete or incomplete. Electro-

myographic studies and studies of chronaxy are also useful in determining the state of the neuromuscular unit. These methods of testing and their application in nerve lesions are discussed fully on page 24 through 35 in Chapter III and need only be mentioned here.

Tinel's sign, when properly applied, is sometimes helpful in evaluation of complete nerve lesions where suture has been done, or in unsutured cases where the lesion is clinically complete. This test is performed by gently tapping the skin over the course of the nerve distal to the lesion. Tapping is continued proximal toward the lesion until a point is reached at which the patient notes a paresthesia in the skin area supplied by the nerve under investigation. Its primary usefulness is to determine whether or not the regenerating axons have crossed the suture line or site of injury in unsutured cases. It cannot be relied upon, however, to prognosticate the final functional outcome. Perhaps of more importance is its usefulness in determining whether or not re-exploration of the nerve should be carried out when the sign is negative after a suitable interval has passed. For instance, if in a high nerve lesion, after a three month interval, Tinel's sign still remains negative, re-exploration of the suture site should probably be made instead of waiting for the complete time interval of expected regeneration for the nerve.

Treatment of Nerve Injuries

Treatment of the Primary Injury

The principal aim of peripheral nerve repair is the accurate approximation of cleanly severed nerve ends without tension. The decision for primary or delayed suture should be determined at the time the wound is first examined. Clean lacerations by sharp instruments are best handled by primary suture, while contused and ragged lacerations with in-driven foreign material, or extensive soft tissue and bone injury, necessitate a delayed suture. If primary suture is decided upon, careful debridement of the wound is performed. Each injured structure, particularly the severed tendons and nerve is identified. This is most easily and most accurately done if the wound is extended vertically both in a proximal and distal direction so that the surgeon can identify normal structures above and below the site of injury. Extending the wound to normal structures avoids such errors as seen particularly in median nerve injury where not too infrequently one end of the severed median nerve has been sutured to the opposite severed end of the palmaris tendon. The extension of the incision should not cross a normal flexion crease. After the ends of the nerve are isolated, a suture may be placed through the nerve in a normal area above and below the injured site. The nerve ends should be cleanly divided with a safety razor blade and bleeding from the stump controlled by placing a small pledget

of gelfoam against it for a few moments. This should be removed before the sutures are placed. The freshly incised nerve ends are not touched with instruments throughout the remainder of the procedure. The through and through sutures are used to handle the nerve ends thereby avoiding damage by instruments. In some instances it may be necessary to further extend the incision in order to obtain more length of the nerve for approximating the ends without tension. If the ends cannot be approximated, the nerve is separated from its normal bed for varying distances so that some traction may be applied in order to effect the union, or the nerve may be given a new bed as in placing the ulnar in the cubital fossa.

However, one must be careful not to destroy or impair the blood supply to the nerve. If the blood supply is impaired through the alignment and *approximation is perfect, function may not be restored.* The nerve ends should be aligned in their proper anatomical position, and, if necessary, a fine epineurial suture can be placed on each segment at a distance from the suture site as a landmark for proper alignment. Suture material does not seem to be as important in nerve regeneration as proper technique in using it. Either .003 inch tantalum wire or 6-0 ophthalmic silk on atraumatic needles may be used. The tantalum is a little more difficult to manage but has the advantage of not "dragging" through the epineurium. For large nerve trunks .005 tantalum, or 5-0 ophthalmic silk, may be used. For the most commonly involved nerves, such as the median and ulnar, a sling stitch through the center of the upper and lower segments is not necessary. It may, however, be helpful if suture of a larger nerve trunk, such as sciatic, is performed, as it helps to approximate the inner nerve bundles. Approximation of the nerve ends is made by placing these sutures through the epineurium avoiding injury to the axons as much as possible. Each suture should be placed sufficiently close to the next one so that there are no gaps in the epineurium through which axons can protrude through the suture line. When the suture is completed, the nerve ends should be gently approximated and the nerve not under tension. In order to avoid tension on the suture line it is frequently necessary to use a molded splint to keep the extremity in a position offering maximum relaxation to the nerve during the healing period. This splint should not produce an angle exceeding 90°. Hemostasis should be secured, divided tendons sutured, and the wound closed with buried fine silk sutures and either silk or nylon sutures in the skin.

Delayed Repair of Nerve Injury

The time for delayed suture is dependent upon several factors. Infection, extensive soft tissue injury, and fracture, may cause delay in the time interval between injury and secondary nerve suture. The optimum time

for suture depends upon the condition of the wound, but it should be emphasized that for best results, the sutures should be carried out within a period of three months from the time of injury. In many instances, delayed suture can be carried out as soon as the primary wound heals and has been demonstrated to be free of infection. This purposeful delay in contused wounds gives an adequate time interval to determine more accurately the extent of damage and intraneural fibrosis in the proximal segment of the contused nerve, and permits a more accurate guide as to the exact amount of nerve end to be resected to reach normal axons. It must also be emphasized that during the delay period, while waiting for tissue healing to occur, the muscles should be kept in optimum condition by appropriate physical therapy. Surgical technique in delayed suture is essentially the same as that for primary suture. The extremity must be widely prepared in anticipation of more extensive mobilization of nerve segments due to necessary resection of scar tissue at the nerve ends. The incisions must be planned to avoid the normal flexion creases and also planned to conform to the scar of the previous incision resecting it when possible. The nerve is approached through normal tissue, if possible, and dissection carried along the nerve to the site of injury. If the nerve is found to be completely sectioned, or has a neuroma in continuity which does not permit the passage of electrical impulses, an appropriate length of scarred nerve ends is resected and suture is accomplished, using the same technique as in primary suture. It might be added that with delay the epineurium is usually more firm and suturing more easily accomplished if the gap is not too great.

Frequently the injury producing nerve injury is a closed one such as fracture or contusion and there is doubt that complete anatomical severance of the nerve exists. In such cases spontaneous regeneration is possible and, in fact, may be superior to nerve suture. In such cases considerable judgment must be exercised in determining whether exploration of the nerve is advisable. If the nerve injury is a contusion, rapid return of function may occur. If return of function is not apparent in the first few weeks, exploration of the nerve may be advisable to obtain a repair during the optimum first three month period. It is in these patients that nerve stimulation and electromyographic studies may be of considerable help. If, after two to three weeks, the muscles supplied by the involved nerve show evidence of denervation by electromyographic studies, and no response to electrical stimulation, early nerve exploration is indicated. If, on the other hand, these studies show evidence of partial function, one should be conservative in recommending exploration, as statistical evidence shows that those patients showing early partial function through nerve injury go on to a recovery more satisfactory than results that can be obtained by nerve suture. An accurate history as to the nature of the initial injury pro-

ducing the nerve lesion may also help in deciding whether or not exploration of the affected nerve is indicated. Stretch injuries of the brachial plexus, sciatic nerve and peroneal nerves produce intrinsic axonal injury which is usually too extensive to permit surgical resection and repair of the involved nerves and is usually not helped by surgery.

Nerve Regeneration and Nerve Grafts

Following either primary or delayed suture, frequent periodic examinations should be performed to determine the progress of nerve regeneration. Many studies have been made on the rate of regeneration of sutured and injured nonsutured nerves. For practical purposes, a rate of regeneration of 1 millimeter per day, or approximately 1 inch per month, is a satisfactory one to follow. This may be altered considerably by loss of blood supply to the nerves through extensive mobilization, a factor that must be considered when anticipating time for a functional return. Tinel's sign is of some value in following the progress of regeneration across a suture line or from the site of a nonsutured nerve injury, as it gives evidence of progressive advance of regenerating axons. A more important clinical feature is the knowledge of the distance from site of suture, or injury, to the first muscle supplied by the nerve. It is in this period where electromyographic studies may again be helpful in determining the presence of regeneration as electromyographic evidence of regeneration, such as disappearance of fibrillation, and the appearance of motor units is present before clinical evidence of muscle function becomes apparent. It is only by these repeated examinations that decision can be made for re-exploration if regeneration is not progressing satisfactorily.

If the nerve is re-explored, decision for further treatment is usually determined by inspection and palpation of the nerve, stimulation of it, and injection of saline through a fine hypodermic needle introduced into the nerve trunk. A combination of these maneuvers usually determines the extent of fibrosis present within the nerve and enables one to make a decision as to whether or not complete resection of the neuroma and reanastomosis is indicated. When resecting neuromata, it is wise to bear in mind the critical length which might be resected and adequate regeneration still obtained. In experienced hands, in adults, these lengths are approximately 7 cm. for the median nerve, 10 cm. for the ulnar nerve, 7 cm. for the radial, 9 cm. for the peroneal, and 7 cm. for the tibial In children, these lengths are probably less. Whenever possible, however, all attempts should be made to mobilize the nerve as better results are obtained in doing this than can be obtained either with nerve grafts or with regeneration through scarred nerve ends.

In instances where it is impossible to overcome a long gap in a divided

nerve by mobilization and positioning, it may be necessary to resort to the use of a free graft to approximate the nerve ends. At the present time the only satisfactory grafting material is autogenous nerve. Because of this, considerable judgment must be exercised in determining whether the functional result to be expected by grafting, justifies the added deficit made by sacrificing the nerve to be used as a graft. The nerves available for grafting are usually cutaneous sensory, such as the internal cutaneous of the forearm or sural. Occasionally major nerve trunks, such as ulnar or radial, may be used when a combination of median and ulnar, or median and radial injury occurs, and the necessity of accurate median repair justifies the sacrifice of the ulnar or radial nerve. Since the diameter of the graft must be equal to or exceed the diameter of the distal stump, it is usually necessary to use multiple or cable grafts when a cutaneous nerve is the source material. The many technical details of making a nerve graft successful might be summarized briefly, as follows: (1) The time interval between injury and repair should be as brief as possible after elimination of the possibility of sepsis; (2) the graft should be placed in as near normal tissue bed as possible to insure vascularization; (3) the proximal and distal neuromata should be entirely removed; (4) the graft should be as short as is consistent with suture without tension, and (5) Pedicle grafting to preserve blood supply may be utilized, if the gap is long.

Splinting of Paralyzed Muscles and Peripheral Nerve Injuries

As a general principal in nerve injuries, splinting, particularly over a prolonged period of time, should be avoided. At times it is necessary to use temporary splints to posture the extremity in the optimum position and to relax tension on the suture line, or because of extensive soft tissue injury or fracture. When such splinting is necessary, the wrist and fingers should be left free, when possible, to permit a physical therapist to mobilize the joints. Intermittent splinting should be used in radial nerve palsy with wrist drop and peroneal nerve palsy with foot drop. These splints should be arranged so that they can be removed with facility to permit passive movement of the joints to be carried out several times daily.

Physiotherapy in Peripheral Nerve Injuries

Physiotherapy should be started as soon as possible following a nerve injury, even if it is necessary for the extremity to be mobilized temporarily in a splint during the period of healing. It is often possible to arrange this splint in such a manner that at least the distal musculature can be massaged and joints moved through their normal range of motion. Passive manipulation by the patient and the physical therapist is extremely important during the time interval between suture and the period of begin-

ning regeneration. Electrical stimulation of the denervated muscles has not proven to be of sufficient benefit during this time to be practical. A possible exception to this is its use in the muscles of the hand and face where it has been shown that atrophy is less in electrically stimulated muscles than in those not stimulated. Electrical stimulation is also useful when muscle function is returning. In this instance, it may help considerably in the re-education of reinnervated muscles. It is important that the patient be taught the principles of self-physiotherapy and occupational therapy. These should be directed along the lines of performing every day activities.

Results of Nerve Suture

In children, primary suture of cleanly lacerated nerves at the wrist, both median and ulnar, often produces exceptionally good functional results. Good results are also obtained following injury to the radial nerve. Poorest results are obtained in injuries of the brachial plexus, sciatic, and peroneal nerves.

Tendon Transplants to Overcome Functional Deficit

When all effort has failed to produce adequate return of muscle function following proper nerve suture, transplantation of various tendons may be in order to provide a functional extremity. The technical details of tendon transplant is beyond the scope of this chapter. It is sufficient to say that it is most generally used in median nerve injuries where opposition of the thumb and fingers is necessary, in radial nerve injuries where extension of the wrist is necessary in order to permit satisfactory grasping, and peroneal nerve injuries to correct foot drop. This subject will be discussed again along with arthrodesis to aid in overcoming functional deficits in Chapter VI on Correlative Plastic Surgery.

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CHAPTER XIX

Surgery of Involuntary Movement Disorders

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FOR THE PAST CENTURY involuntary movement disorders in children have been conventionally referred to as extrapyramidal diseases. This term evolved from the fact that these involuntary motor manifestations were believed to derive from derangement of neurologic mechanisms located outside of or "extra" to the pyramidal tract. In recent years it has become apparent that this inclusive term of extrapyramidal diseases is not an entirely satisfactory one for two reasons:

1. The exact anatomic and physiologic limitations of the so-called pyramidal and extrapyramidal systems cannot be clearly delineated and, in fact, the classification of these two symptoms as separate anatomic or physiologic entities is now considered to be an oversimplification.
2. The term extrapyramidal disease has come to cover a confusing host of widespread manifestations of neurologic abnormalities such as increased or abnormal motor movement, decreased motor movement, increase or decrease in muscular tone, as well as a host of auxiliary symptoms such as autonomic and vegetative phenomena, which often appear in the course of the so-called extrapyramidal disorders. For that reason there has been a welcome tendency in recent years to attempt to delineate each of the so-called extrapyramidal disorders by more precise clinical or anatomico-physiologic denotation.

It is the purpose of this chapter to deal with those manifestations of "extrapyramidal disorders" in children which can best be described as involuntary movement disorders, this term being used to denote abnormal increase of muscular tone as well as involuntary movements. Therefore, these clinical entities will be approached on the basis of individual signs and symptoms rather than in the conventional manner of descriptive nosologic disease states. It should be clearly pointed out that a consideration of the neurosurgical alleviation of extrapyramidal disorders really signifies only a consideration of the neurosurgical treatment of certain symptoms,

usually those of the involuntary movements and abnormally increased muscle tones produced by these disorders.

Involuntary Movement Disorders Which May Be Treated Neurosurgically

There are a large group of hyperkinetic phenomena in which surgical therapeutic possibilities exist, some of which have already proven to be fruitful. Anatomic and physiologic investigation in the human have not yet reached the point which will enable us to present a comprehensive analysis of each movement disorder in relation to the particular neuronal aggregate or circuit which is responsible for that particular disorder and therefore such an analysis will not be attempted in this chapter.

The etiology of most of these various clinical entities is for the most part unknown and except where otherwise indicated the neurosurgical approach to relief of various hyperkinetic phenomena will be the same regardless of the etiology of the particular involuntary movement. For example, rigidity, tremor and dystonia can be best treated by a specific destructive lesion of the globus pallidus and/or thalamus regardless of whether the symptoms are on a postencephalitic or a so-called idiopathic basis. It is for this reason that a detailed discussion of etiology need not concern us in this chapter.

Rigidity

Rigidity may be described as an abnormal increase in the tone of voluntary muscle. Such an increase in muscular tone may be considered to be abnormal when it begins to interfere with voluntary movement or involuntary spontaneous, automatic movements of these muscle groups. Abnormal increase in muscular tone may be present in the extremities, the trunk, or nuchal musculature and may be reflected in hypokinesia, bradykinesia, or abnormal postures of the various parts of the body. In some instances this rigidity may cause abnormal forced postures which when present for long periods of time may result in relatively fixed contractures.

Rigidity must be clinically differentiated from the hyperkinetic state referred to as spasticity. The latter is almost invariably accompanied by the phenomenon of clonus and by a positive Babinski sign. These phenomena are not present in cases of rigidity alone. Moreover, passive movement of a rigid extremity results in regular periodic resistance to such movement which is referred to as cogwheelism, whereas passive movement of a spastic extremity yields a uniform resistance which gives way suddenly resulting in the so-called clasp-knife phenomenon.

Rigidity is commonly encountered in children in postencephalitic states and is sometimes present in children who present abnormal neurologic

signs as a result of birth trauma. This latter group usually falls within the non-specific classification of cerebral palsy.

Rigidity and rigid fixed postures can be successfully alleviated by the placement of a large destructive lesion in the globus pallidus opposite to the side of the rigid extremities. Placement of such a lesion may be carried out in numerous ways, and these will be described further under the section of this chapter on surgical technique. Of all of the involuntary movement disorders, rigidity may be considered the most readily amenable to surgical therapy at the present time. Neurosurgical therapy or, more specifically, surgery of the globus pallidus, is the treatment of choice of incapacitating rigidity.

Tremor

Tremor is more frequently seen in adults than in children, it being a relatively rare sequela of encephalitis in children. The type of tremor which is considered to be an "extrapyramidal" phenomenon is an alternating tremor at rest. That is, it is characterized by rhythmic purposeless contractions of antagonistic muscles, the average rate of these antagonistic contractions being from 4 to 8 per second. Such tremor is usually not accentuated by active muscular movement and, in fact, may temporarily disappear during purposeful motion of the involved muscle groups. This tremor disappears during sleep and is usually increased by emotional duress and excitement. When present in the fingers it sometimes assumes the so-called pill-rolling attitude so frequently seen in paralysis agitans. Alternating tremor may involve the fingers, the hand, or the entire upper extremity. Similarly, it may involve only the distal portion of the lower extremities or may become major tremor involving the entire limb. Alternating tremor of the lips, facial musculature and nuchal musculature are not uncommon. However, the typical alternating tremor at rest involves principally the extremities and is most disabling when it involves the upper extremities thus interfering with fine motions of the fingers.

Alternating tremor has been the focal point of neurosurgical investigation for many years. Several techniques intended to interrupt the tremor mechanism at different levels have been employed with varying degrees of success. Most of these techniques were planned as an attack upon the pyramidal pathways or those "extrapyramidal" fibers arising from the premotor cortex. In all these operations the degree of lasting relief of tremor was proportional to the sacrifice of motor power. Relief of tremor without production of motor weakness or paralysis has been achieved by operation upon different structures included in the concept of extrapyramidal system. These operations include Meyers' and Fenelon's open techniques, several

types of stereotaxic methods and our own techniques of chemopallidectomy and chemothalamectomy which will be discussed later in this chapter.

Chorea

Chorea, in common with the other involuntary movement disorders, can be produced by various etiologic agents. Choreic movements in children may develop during the course of febrile illnesses and they are particularly common as one of the manifestations of rheumatic fever. The involuntary movements of chorea are more rapid and purposeless than those of tremor or athetosis. These rapid irregular purposeless movements are most marked in the upper extremities but may affect the face, the trunk, the nuchal musculature and the lower extremities as well.





FIGURE 1. *(Left)* A bilateral congenital athetosis demonstrating a fairly typical athetotic hand. In this case the athetosis is superimposed over a double hemiplegia.

FIGURE 2. *(Right)* This case demonstrates dystonia musculorum deformans. Note the fixed postures and deformities. However, there is no paralysis or pyramidal tract involvement underlying the involuntary movement disorder in this case.

The choreiform movements accompanying infectious diseases, the most common form of which is Sydenham's Chorea, are usually self-limited manifestations and do not require surgical therapy.

Athetosis

Athetosis is one of the more gross, violent, abnormal movements of voluntary musculature. It is a slow twisting vermicular or writhing type of movement involving principally the extremities but also causing deforming writhing movements of the trunk, head and neck. At times these slow vermicular movements may be combined with sudden irregular jerky choreiform movements, the combination usually being referred to as choreoathetosis.

The most common cause of athetosis is birth injury which causes widespread damage to the intracranial structures particularly the basal ganglia. The manifestation of athetosis has been most commonly ascribed to lesions in the caudate nucleus and putamen. Although there is some evidence for this, it cannot yet be considered certain that these lesions alone are productive of athetosis.

Athetosis as a pure involuntary movement disorder is rare. It is most often combined with signs of the pyramidal tract diseases plus impairment of other brain functions so that children suffering from unilateral or, more commonly, bilateral or double athetosis frequently have this athetosis superimposed on a spastic paresis of the extremities. Furthermore, they are usually disabled by marked speech defects and mental deficiency. Therefore, children demonstrating athetosis as part of an overall symptom complex consisting of mental deficiency, speech retardation, spastic paresis of the extremities, and involuntary movement disorders, must be considered as less favorable candidates for surgery than those children who demonstrate involuntary movements uncomplicated by spastic paralysis or mental deficiency. Nevertheless, athetosis as a symptom can be favorably treated by neurosurgical means.

Dystonia

Dystonic movements are slow, long sustained twisting or turning movements which may involve the extremities, the head, the neck and the trunk. These movements appear to be slower and even grosser than the movements of athetosis. They may gradually become sustained so that the limb or a part of the body remains in a twisted deformed position, and are referred to as dystonic fixed postures. The muscles involved are under great tension or markedly increased tonus during dystonic movements or dystonic postures. In contrast to choreiform motions which are quick and irregular with a minimum of deformity or fixed postures, dystonic motions have a minimum

of quick component and are characterized by their slowness, great muscle tension, and fixed postures.

Although dystonic types of movement and fixed postures may be seen in various mixed type of involuntary movement disorder, the most typical example of dystonia is seen in that disease entity referred to as dystonia musculorum deformans. In our experience this type of involuntary movement disorder has a more typical and pure form than the other types described above. Dystonia musculorum deformans usually has its onset at the age of six to nine years. It begins insidiously with intermittent slow dystonic motions of some member of the hand or the foot, though usually the hand and may be mistakenly diagnosed in the former instance, as writers' cramp. At the onset it is usually considered to be a nervous tic or habit spasm. In almost every instance as the disease progresses from a simple twisting movement of one hand or foot to involve the remainder of the extremity or the body and assumes a more grotesque and abnormal appearance, a diagnosis of hysteria is entertained. Soon, however, despite psychotherapy and other conservative measures, the relentless progress of this disease resulting in a deformed helpless child makes the diagnosis of dystonia musculorum deformans inevitable.

The etiology of this disease is unknown. In many instances, however, several cases are seen in various branches of one family. In three of the children who have been treated on our own service within the past year for advanced dystonia musculorum deformans, the mother was found to harbor a lesser form of dystonia, suggesting a familial tendency.

This disease has a relentlessly progressive course resulting in complete incapacitation and helplessness. The movements and deformities become so severe that joints, particularly the hips, may become chronically dislocated and in some instances fractures result from the deforming twisting movements of the extremities. Prior to recent years there has been no satisfactory treatment for this disease.

During the past two and one-half years, ten cases of dystonia musculorum deformans have been operated by chemopallidectomy or chemopallidectomy plus a lesion in the ventral lateral nucleus of the thalamus. Eight of these cases have been relieved of their dystonic manifestations without inflicting any motor or sensory loss on the patient. The earliest of these cases have now been followed for more than 2 years. Two of the early cases in our series of musculorum deformans will be reported below to demonstrate the dramatic response that can be obtained in this particular type of involuntary movement disorder. In our experience, dystonia, being a pure involuntary movement disorder without intellectual deterioration and without concomitant pyramidal tract involvement, is particularly accessible to chemopallidectomy and thalamectomy.

Diagnostic Workup

Diagnosis of an involuntary movement disorder is usually not a problem. Although one may not be able to elucidate the etiology or pathogenesis of a particular involuntary movement, these symptoms are self-evident and it is not necessary, as a rule, to carry out a detailed diagnostic workup in order to arrive at a diagnosis of athetosis or of dystonia. On the other hand a detailed evaluation is necessary in order to decide whether or not a particular patient would be a suitable candidate for neurosurgical intervention.

The aim of the workup of a child with involuntary movement disorder is to enable the surgeon to decide whether or not successful alleviation of the involuntary movements by surgical means will benefit the patient enough to warrant surgical intervention. For example, if a congenital athetotic patient is so mentally retarded that relief of the involuntary movements alone will in no way aid the nursing care, contribute to the comfort of the patient, or make that patient a candidate for rehabilitation, then obviously surgery aimed at the involuntary movements alone is not worthwhile. If, on the other hand, relief of the involuntary movements will enable the particular patient to be rehabilitated, or render him more comfortable, or will make nursing care easier and more profitable then surgical intervention for involuntary movements may be considered. In order to evaluate candidates in this category for neurosurgical treatment we believe that one full week of hospitalization is mandatory. During this evaluation in addition to the detailed history and neurologic examination each patient is subjected to a complete medical workup, psychological evaluation, speech evaluation, muscle examination by the department of rehabilitation, and ancillary tests such as electroencephalography and electromyography.

History

The method of taking a detailed history obviously need not be reviewed here. However, emphasis on certain points in the history can be referred to as an aid in the eventual selection of patients for surgery. Ordinarily at least part of the history is taken from the parents. It is important during this part of the history taking to insist that the parent outline what has been the principle incapacitation of the child during the development of his illness. If the parent is preoccupied with the mental retardation, or with elements of the disease which are due to pyramidal tract involvement, this must be carefully borne in mind when a decision is made as to whether the patient should be subjected to surgery. If on the other hand, during the history taking, it is obvious that the involuntary movements are the principle source of distress to the patient and his surroundings, then this will have a very positive effect in the ultimate evaluation of the patient.

Neurologic Examination

Although one seldom needs a detailed neurologic examination to arrive at a fairly accurate diagnosis of a particular involuntary movement disorder, it is most essential that an extremely meticulous neurologic examination be performed in each of these cases if they are to be considered as candidates for neurosurgery. One must lay particular emphasis during the course of this examination on the conscious level of the patient, his orientation and ability to cooperate. These particular facilities of the patient will play an important role during the postoperative course and during rehabilitation. Speech must be very carefully evaluated and disabilities of speech must be interpreted so that one may postulate whether they are amenable to relief of rigidity of the facial musculatures or whether such defects are due to cortical involvement or mental retardation.

The degree of voluntary motor movement that underlies the involuntary movements must be very carefully appraised. Present neurosurgical techniques are aimed at relieving involuntary motion. Some of these techniques are capable of relieving involuntary motion without inflicting even a temporary weakness or paralysis on the patient. However, if such weakness or paralysis exists preoperatively it must be evaluated, recorded, and explained to the parents prior to surgery that the aim of the surgical technique is to relieve involuntary movements and that such techniques are not capable of producing voluntary movements which do not exist by reason of preoperative pyramidal tract lesions. On the other hand, voluntary movements which are merely suppressed by superimposed rigidity or by excessive involuntary movements may be markedly improved during the postoperative period. Spasticity accompanied by clonus and positive Babinski sign should be carefully differentiated from rigidity.

Medical Workup

An extensive medical workup is carried out on each patient who is a possible candidate for neurosurgical treatment of involuntary movements. Among the more important conditions that are ruled out during the course of this workup are electrolyte imbalances, abnormalities of temperature control, and other neuro vegetative phenomena which are not uncommon in this group of patients, particularly in the congenital choreo-athetotic patient. Unless such abnormalities can be corrected preoperatively they are considered to contraindicate neurosurgical intervention.

Psychologic Evaluation

One of the most important parts of the preoperative study is a detailed psychologic evaluation which is considered to be imperative not only in

evaluating children but also adults as possible candidates for surgical treatment of involuntary movements. Evidence of organic brain deterioration due to cortical involvement or other brain lesions contraindicate basal ganglia surgery. We have found that children and adults with average or superior intelligence suffer no intellectual impairment from this type of neurosurgical intervention. On the other hand such patients who have advanced organic mental changes and intellectual impairment may be more confused or disoriented postoperatively. In addition it is the aim of our psychologic evaluation to try to determine the patient's ability to react to the operative and postoperative stress and to delineate any tendency towards psychotic or abnormal behavior. Extensive psychologic studies are also carried out postoperatively in each of our cases.

Evaluation by the Department of Physical Medicine and Rehabilitation

At least one member of the technical staff and one physician of the department of physical medicine and rehabilitation does an extensive muscle evaluation and carefully studies the activities of daily living of each patient during the preoperative workup. This enables the members of that department to prognosticate the ability of the patient to be rehabilitated in the event that the involuntary movements are successfully alleviated by operation. If this evaluation leads us to believe that rehabilitation will be impossible even if the involuntary movements are successfully alleviated then a decision regarding surgery must be based solely on whether it is indicated in order to make nursing care more feasible and the patient more comfortable. If, on the other hand, we are led to believe that the patient can be successfully rehabilitated in the event that the involuntary movements are relieved, then this knowledge plays an important role in fashioning the decision for operative intervention.

Ancillary Tests

Ancillary tests such as electroencephalography play an important role in the preoperative evaluation. For example, electroencephalography helps to determine the presence of any focal lesion productive of an epileptic tendency which might mitigate against operative intervention. Moreover, the presence of diffuse electroencephalographic abnormalities may tend to strengthen our conviction that the patient had diffuse or generalized brain involvement with mental deterioration and consequently would not be a suitable candidate. Conversely a normal electroencephalogram would obviously be interpreted favorably in evaluating a patient as a surgical candidate. Electromyography plays a useful role particularly in evaluating which muscles are more involved in abnormal movements of the neck. In some instances electromyography is used merely for scientific verification of the

clinical picture. Spinal puncture is rarely necessary as a preoperative test.

Following the workup referred to above an evaluation conference is held which is attended by each of the physicians who contributes to this preoperative evaluation. By this means it is possible to evaluate the patient as a whole and to decide whether or not surgery for the relief of involuntary movements is indicated in a particular case. Since medical therapy up to the present time is unavailing in these cases one must weigh surgical therapy against the natural course of the disease if such therapy is not undertaken. By weighing each of the factors enumerated above and discussing this type of elective surgery with the patient and/or his parents a decision is then reached as to whether surgical intervention should be carried out.

Pathologic Physiology of Involuntary Movements

Before entering into a discussion of surgical treatment of the disorders of involuntary movement, namely: rigidity, tremor, choreo-athetosis and dystonia, we are going to review briefly the basis of the surgical approach in the light of our present knowledge of the pathologic physiology of these disorders.

From a therapeutic standpoint the existence of a motor system which had already developed to a functional degree prior to the onset of the illness, is extremely important. There are patients in whom the disease appeared late in childhood after a presumably normal period and in these patients the relief of the involuntary movements should restore the motor function to its previously "normal" level. There are some other patients who were born with a motor defect or acquired it during early infancy before a voluntary pattern of movements had time to develop. Even if the anatomical lesions in both groups involve similar areas, it seems that from a physiological point of view we are dealing with two different problems. And finally there is a third group of patients in whom the involuntary movements are only a part of the widespread disease of the motor system involving the "extrapyramidal" as well as the "pyramidal" mechanisms of movement.

Pathology of Involuntary Movements

It is unfortunate that our present knowledge of the pathology encountered in patients affected by disorders of involuntary movements cannot be integrated into anatomical, physiological or clinical concepts. As Greenfield has stated recently ". . . in the diseases of the extrapyramidal system . . . there are many problems of etiology and pathogenesis but the most difficult and least understood is that of the relationship of symptomatology to anatomical lesions. . . ."

The interest in the pathology of the extrapyramidal system as related to disorders of involuntary movement dates from 1911 when Alzheimer, Vogt

and Wilson described their findings in cases of chorea, status marmoratus and hepato-lenticular degeneration. From that moment on, such symptoms as rigidity and tremor were explained in terms of rather vague lesions in the striatum and pallidum.

Specific pathologic lesions have been described in some clinical syndromes but we cannot accept today such classical oversimplified concepts as "paralysis agitans is due to lesions of the pallidal system, Huntington's chorea to lesions of the neostriatal system, while the syndromes of Cecil Vogt and Kinnier Wilson are caused by involvement of both systems."

In such complex extrapyramidal syndromes as Parkinson's disease, lesions considered as specific were described by Dana in every area of the neuro-motor system: from the "vessels of the pia mater to the nerve cells of the pons to the connective tissue of the nerves and muscles." Even when the attention of the pathologists became concentrated on the extrapyramidal structures the focus of attention shifted from time to time from the pallidum to the putamen to the locus ceruleus to the substantia nigra. A similar range of variation is found in the description of the actual pathological lesion: Hyaline inclusions and degeneration, neurofibrillar changes, small lacunae, loss of cells, degeneration of the medullary sheath. The lack of consistent findings in a disorder as thoroughly investigated as paralysis agitans is well expressed by Lewey when he states: "When I had examined pathologically the seventh dozen of parkinson brains I was completely confused because you seemed to be able to prove just as well a theory as the contrary one."

Traditionally hemiballismus has been presented as a result of a localized lesion of the corpus Luissi both clinically and experimentally. Hemiballistic movements may however appear with an intact nucleus subthalamicus, the responsible lesions being confined to the neo and/or paleo striatum, the ventral thalamus or the post central gyrus according to Meyers.

Status marmoratus of the striatum, which is characterized by the findings of abnormal myelinated fibers throughout the posterior halves of the putamina, has been considered as the most common etiology of double athetosis. Other lesions of the striatum and/or pallidum as status dysmyelinisatus, and vascular necrotizing lesions, have also been reported in relation to athetosis. Dystonia musculorum deformans is usually related to lesions in the neostriatum especially shrinkage and sclerosis of the putamen. Whether these lesions are the result of birth trauma, infantile encephalitis or hereditary anomaly remains to be elucidated. Marked extrapyramidal symptoms with athetotic and dystonic features have also been found by Carpenter to be associated with status marmoratus of the thalamus. On the other hand status marmoratus of the thalamus has been occasionally found by Norman without extrapyramidal symptoms, and a distinction between patterned and nonpatterned abnormal movements cannot be made solely on pathological basis.

This variety of lesions—both in nature and in localization—associated with syndromes of abnormal movement, prevents the surgeon from planning therapy solely on pathological grounds. We have to limit ourselves to consider the disorders of movement as groups of syndromes related in an unspecific way to anatomical changes within the basal ganglia of the brain. In this manner it is possible to accept—if not to understand—how similar clinical entities may respond to different surgical measures and how identical lesions may have the same favorable effect in different types of clinical manifestations. These considerations will be further emphasized when we discuss the need of a flexible technique and the variable placement of our lesions in equally successful cases.

Our present knowledge of the physiology of the basal ganglia does not allow us to approach the therapy of extrapyramidal disorders on a solid conceptual ground. The results of animal experimentation have been rather controversial and although providing us with useful information they have not contributed a great deal to a rational surgical therapy.

If we accept the fact that the pathological lesions are variable and we cannot yet elucidate a complete anatomo-clinical correlation, then we must consider the clinical manifestations as a result of the disturbance of a complex neuro-physiologic mechanism. We may assume from a theoretical point of view that we are dealing with areas of function rather than semi-isolated structures and that a lesion within the functional boundaries of that area would manifest itself by similar symptoms with independence of the specific structure mainly affected. Moreover, there may be such a thing as a dynamic type of malfunction; in other words, a lesion in an area of the brain may act not only by producing a deficit but also as a disturbing relay in a circuit of impulses. Further, the responsive mechanism set into motion by the abnormal relay may be a different one in cases with similar clinical manifestations or the deranged impulses may act at different levels with different results. This leads us to postulate that the same circuits can be surgically interrupted at different points with similar results, a theory that has been borne out in our experience.

Unfortunately there is not enough physiological information to provide us with an accurate picture of the modus operandi of the different neuronal circuits, or their functional balance and interrelations. The anatomical studies give only limited information regarding the nonmyelinated fibers connecting the structures of the diencephalon and mesencephalon and projecting into the cortex and the basal ganglia. The integrative action of the thalamo-reticular system, the organization of the projection systems, the thalamo-pallidal connections, the response movements and action potentials induced in the cortex by reticular, hypothalamic and pallidal stimulation are problems under active study by Papez, Magoun, Dusser de Barenne, and

Russell. It remains to be known how much of the data gathered by means of such techniques as neuronography and electrode implantation and stimulation should be considered as physiological. At the same time Meyers and Lassek inquired into the validity of such concepts as extrapyramidal and pyramidal systems in search for a more dynamic, integrative understanding of motor function.

From a practical point of view we have found it useful to classify patients in the three categories previously mentioned: (1) extrapyramidal syndromes appearing after a period of "normal" motor function. (2) Extrapyramidal syndromes in children who never developed a motor function of voluntary organized use, including the congenital cases and those on whom the lesion occurred at the time of birth or shortly thereafter. (3) Syndromes with mixed pyramidal and extrapyramidal symptomatology. This classification is independent of the known or suspected etiologic agent.

In the first group of patients the relief of the involuntary movements restores the motor function to its previous normal level. In the second group we have to count on the postoperative rehabilitation potentials to develop such a voluntary system. In the third group of patients the surgical goal is a limited one. Whatever is achieved in these groups of children will depend both on the surgical procedure and the hidden capabilities of that particular brain.

Surgical Techniques and Results

"On the whole surgery has little application in this vast field . . . of the extrapyramidal hyperkinesias."

These words of Bailey quoted above summarize the feelings of neurologists and neurosurgeons when evaluating the results obtained by the techniques introduced up to 1951 for the surgical relief of involuntary movements. These operations belong to three main categories: open attack upon the pyramidal pathways at all levels from the precentral motor cortex to the spinal cord. Oliver felt that all of them, as well as pedunculotomy introduced at a later date, "seemed to boil down to artificial production of paralysis." A second group of techniques were developed which were able to demonstrate that relief of rigidity could be obtained by a direct section of the ansa lenticularis, thus diverting the attention from the pyramidal to the extrapyramidal structures. A third group of operations resulted as a combination of Meyers' demonstrations of the importance of the pallidofugal fibers in the pathogenesis of rigidity and of the development of stereotaxic techniques for humans by Spiegel and Wycis. There are several variations of the stereotaxic approach, all of them using basically the system of coordinates of Horsley-Clark and a lesion was produced by the passing of an electric current by Spiegel and Wycis, thermo-coagulation by Leskell, or procaine-wax injection by Narabayashi.

In general terms it can be said that operations upon the pyramidal tract had to pay the price of a paralysis in order to relieve certain manifestations as tremor and they were not effective in the relief of rigidity. The pure stereotaxic methods with the globus pallidus and the ansa lenticularis as surgical targets have limited application due to technical difficulties and have obtained some success in the relief of rigidity but have not produced large enough lesions in most instances to permanently relieve tremor and gross involuntary movements.

Following our early experience with anterior choroidal artery ligation, another method of pallidal destruction by progressive chemical neurolysis was described in 1953. This technique was intended as a simple practical way of attack upon the components of the extrapyramidal system, and did not make use of a system of coordinates but relied on clinical testing for the placement of the neurolytic lesion. This operation was basically conceived on the assumption that the ultimate placement of the lesion should depend on the clinical results and not solely on the radiological position of the needle. The variability of the anatomical lesion underlying the extrapyramidal disorders of movement and the lack of physiological and clinical correlations have already been discussed. Another important factor is the range of variations of the measurements of the brain so that precise stereotaxic procedures may give to the surgeon a sense of security without actually leading him to the anatomical structure that he had in mind. Therefore, every kind of therapy must submit itself to the clinical test of the results and this has to be the criteria for the selection of a certain procedure no matter how puzzling the anatomical and physiological complications might be.

The surgical technique aiming at the treatment of involuntary movement disorders should ideally have:

(1) A physiological purpose so that the relief of a disorder is not achieved at the expense of a normal neurological function.

(2) Enough flexibility to cope with the problem of the different pathogenesis of the disorders.

(3) Enough safety and accuracy so as to permit the placement of predictable lesions within the intended limits of anatomical structures.

(4) A physiologic test or method of placing a reversible lesion prior to infliction of a permanent destructive intracerebral lesion.

(5) Ability to produce a lesion of sufficient size to permanently relieve symptoms without inflicting any neurologic deficit.

If these premises are observed, physiological and anatomical data may be gathered through such a *technique in order to establish* solid foundations for further procedures. We cannot a priori determine what the clinical response to a certain lesion is going to be based on the information received from the laboratory since laboratory animals do not have these diseases.

What we know about surgical treatment of extrapyramidal disorders comes from the observation of patients following the surgery upon the pyramidal tract, the open attack upon the extrapyramidal structures, and the more recent methods of selective intracerebral lesions.

We now should consider the actual technique of some of these procedures and what we may expect from them in clinical results and in neurological consequences. The indications, technique and results of our methods of chemical neurolytic lesions into the globus and the thalamus by means of an indwelling catheter will be discussed in detail.

Stereo-Encephalotomy

Spiegel and Wycis have developed a technique modifying the Horsley Clark technique for animals which consists essentially in the intracerebral placement of an electrode according to a pre-established reference point in a system of coordinates by an instrument rigidly attached to the skull. The position of the electrode is determined by roentgenographic control and an electrolytic lesion made by passing a direct current through the electrode. The anodic lesion thus produced would have a diameter of 2-3 mm.

The published results of Spiegel and Wycis are evaluated in terms of diminished tremor or voluntary movement and include a series of four patients with Huntington's chorea, one case of electric chorea and seventeen cases of resting tremor. Of the four patients with Huntington's chorea two were definitely improved, one was moderately improved in one side and the other patient developed a hemiplegia. Of the seventeen patients with resting tremor, a lesion in the ansa diminished the tremor in one of them and there was one postoperative death. About 50% of the patients had some degree of improvement in rigidity and bradykinesia.

Results similar to those reported by Spiegel and Wycis have been reported following the techniques of Reichert, Talairach, Guiot and Brion, all of them using electro-coagulation aimed at the ansa lenticularis. The reports concern mainly patients with Parkinson's disease and the degree of reported improvement without a specific distinction between tremor and rigidity varies between 60 and 70% on the operated cases. Some of the reports are not easy to evaluate because there is no clear definition of what is meant by improvement. Because some of the failures of the electro-coagulation seem to be due to the production of lesions not large enough, two electrodes have occasionally been introduced so as to make the electric current pass between both of them thus creating a double lesion. Lexsell has claimed that with bipolar thermo-coagulation a lesion 1 cm. in diameter could be created.

The stereotaxic operation can achieve a high degree of accuracy in placement of the electrode. It has considerable advantage in anatomic and physi-

ologic studies. However, as practical neurosurgical clinical techniques, the stereotaxic methods have the disadvantage of being time-consuming, involving the need of a complicated and often cumbersome apparatus. Moreover, the results so far reported are not as complete as those achieved by chemopallidectomy.

Ansotomy

Two methods of open ansotomy have been described by Meyers and by Fenelon. Meyers uses a cortical incision and transventricular approach to the globus pallidus. Although Meyers' contribution to the understanding of extrapyramidal disorders was a notable one, this type of operation is not recommended by the author for general usage due to a relatively high mortality rate (15.7%) and postoperative complications, such as convulsions, and inconsistency of clinical results. Fenelon and Guiot have attempted a direct approach to the ansa lenticularis and mesial globus pallidus via subtemporal or subfrontal approaches. Once the intended area has been reached they proceed to pass a coagulating current. Among forty patients operated by Fenelon there were thirty-two improvements, one postoperative death and six patients remained unchanged. Guiot and Brion have reported forty-seven cases of them, forty parkinsonians and seven different types of extrapyramidal disorders. Twenty-six of the forty parkinson patients improved, three died and nine remained unchanged. Of the seven "varied extrapyramidal disorders" three improved, four unchanged and there were no postoperative deaths. There is no specific mention of involuntary movement in children but Guiot states that one must be cautious in the treatment of choreoathetosis.

Cortical and Pyramidal Operations

Cortical procedures were introduced by Bucy and Klemme in an attempt to relieve the tremor of the contralateral extremities. Bucy's procedure involved extirpation of Brodmann area 4 and 6 of the cerebral cortex. This produced a contralateral hemiplegia which subsequently lessened in severity, leaving the patient with a residual hemiparesis but with a reduction or absence of tremor. Some time later Meyers theorized that the undercutting of area 4 might be effective in relieving hemiballistic movements. He carried on undercutting from areas 4S and 6 in two patients with marked improvement in one and a postoperative mortality in the other. Walker in 1957 stated that operations designed to cut fibers from certain precentral zones "have been based upon the assumption that there is a suppressor strip anterior to the motor cortex, a concept which has not been proved for man and for which considerable doubt has been expressed." None of these cortical operations gave consistent results, and are not recommended for clinical use.

(2) The effective posterior or thalamic lesion falls within the nucleus ventralis lateralis of the thalamus and in some cases the postero-medialis as well (Figure 3).

The projection of these areas in the standard AP and lateral x-ray films of the skull taken at thirty-six inches is shown in Figure 3. For our measurements in the lateral view we have used the foramen of Monro and the pineal gland as reference points and the measurements are expressed in terms of a relative position of the lesion over the foramen-pineal line. In view of the fact that the distance from the foramen to the pineal varied in cadavers from 19 to 32 mm. (and that area includes the basal ganglia) we thought that a single reference point would not be reliable. Using a relative position over a variable distance the localization in the cadaver has been accurate and the variations of shape and size of the head in the sagittal plane are auto-

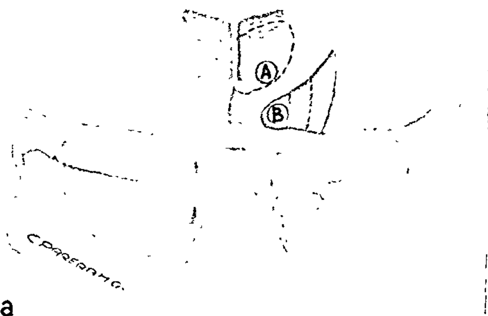


FIGURE 3 (a). A diagramatic representation of the anteroposterior view of the ventricular system demonstrating the two lesions we are now using in these disorders. Site A demonstrates our typical lesion in the ventrolateral nucleus of the thalamus. Site B represents the *typical globus pallidus lesion.*

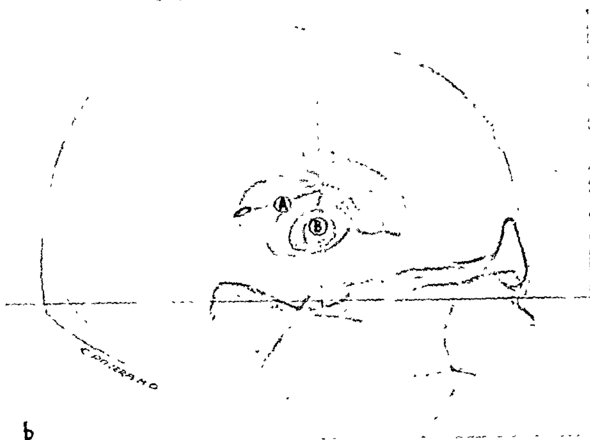


FIGURE 3 (b). A diagrammatic representation of the lateral view of the ventricular system demonstrating the two lesions we are now using in these disorders. Site A demonstrates our typical lesion in the ventrolateral nucleus of the thalmus. Site B represents the typical globus pallidus lesion.

matically corrected. When the pineal is not calcified—as is frequently the case with children—the cisterna ambiens or the recessus suprapinealis should be filled with air and used as a posterior landmark.

In children we have performed these procedures under general endotracheal anesthesia. However, in dealing with parkinsonian adults there is a definite advantage in having the full cooperation of the patient and therefore the operation is carried out under local anesthesia and light pentothal in such a way that the patient is awake by the time of the actual insertion of the needle. This permits clinical testing of the patient throughout the procedure. Due to the violent character of the movements this method of controlled surgery cannot be used in children. Usually we limit ourselves to the placement of the needle and leave the clinical testing of the inflated balloon until the child is awake and neurological testing can be carried out.

The technique of chemopallidectomy-thalamectomy is as follows:

STEP 1—A wire landmark is taped to the skull in the estimated plane of the foramen of Monro.

STEP 2—Pneumoencephalogram with 40 cc. of air enough to visualize the anterior half of the ventricular system is performed. Care should be taken in the identification of the foramen of Monro. If the pineal gland is not

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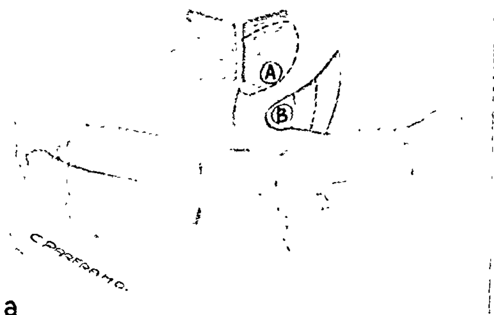


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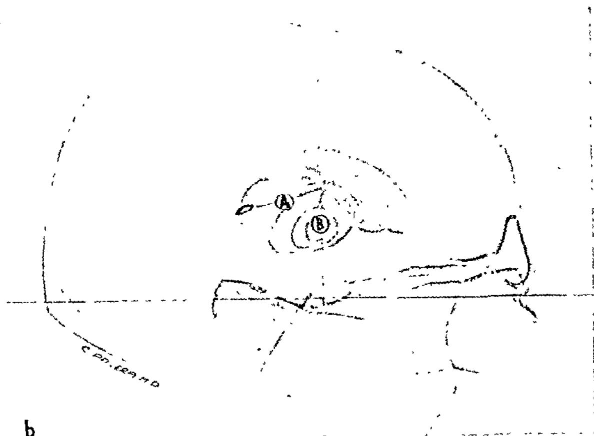


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calcified the cisterna ambiens should also be identified as an auxiliary landmark. The coronal plane landmark represented by the wire is moved when necessary so as to make it coincide with the vertical plane to the base line of the skull passing through the posterior margin of the foramen.

STEP 3—Trephination is carried out over the marked foramen plane at a distance 5-6 cm. from the midline.

STEP 4—Placement of the needle guide. Three pointed screws hold the guide in the desired plane. The needle carrier is poised over the trephine opening.

STEP 5—Insertion of the cannula along the needle carrier. The needle carrier may be moved back and forth and also its angle of attack may be easily changed when a correction in the direction of the needle becomes necessary.

STEP 6—The needle is aimed visually towards the desired point and introduced into the cortex up to a depth of 2-3 cm. The target area has been previously marked in the pneumoencephalogram films in relation to the standard landmarks, so that we know what is the estimated angle that the needle should make with the vertical foramen plane. In the AP view it is directed towards the internal canthus of the eye or slightly more medially depending on the relative size of the ventricles and on the shape of that particular head.



FIGURE 4 (a). Lateral view. X-rays showing the target area in which the balloon at the tip of the needle has been inflated with ¼ cc. of Hypaque.



b

FIGURE 4 (b). AP view. X-rays showing the target area in which the balloon at the tip of the needle has been inflated with ¼ cc. of Hypaque.

STEP 7—AP and lateral x-ray films of the skull are taken. The direction of the needle in both coronal and sagittal planes is thus visualized before introducing the needle to the depth of the brain and the proper adjustments can easily be made by comparing the virtual end point of the needle with the target point as marked in the pre-operative encephalogram. In order to speed up the surgical act, all the x-rays in the operating room are taken on polaroid film which is developed and dried in fifty seconds. With this technique, as many adjustments as necessary can be made rapidly as the needle progresses into the depth of the brain.

STEP 8—Once the target area has been reached, the balloon at the tip of the needle is inflated with ¼ cc. of radiological contrast material (Hypaque) (Figure 4). The distention of the balloon is immediately followed by a relief of the tremor and/or rigidity if the position of the needle is correct. Sometimes the simple concussion effect of the tip of the needle is to alleviate the symptoms for a brief period of time and sometimes there is

a thirty to sixty second latency between the inflation of the balloon and the clinical response. In any case, the response, when there is any, is clear enough both from an objective and from a subjective point of view.

The first function of the balloon therefore is to serve as a clinical test at the time of surgery. If the cannula has been misplaced into the internal capsule, distention of the balloon is immediately followed by contralateral weakness and with return of the motor power when the balloon is deflated. It is possible by rotating the needle to blow up the balloon in different planes and this gives an added flexibility to the procedure.

STEP 9—After the desired result by balloon inflation has been obtained the cannula is fixed to the galea by means of catgut suture and fixed to the skin by means of silk stitches. The balloon is left inflated in place. Keeping the balloon inflated for twenty-four to forty-eight hours before any injection is made gives us two additional advantages: 1) The particular reactions of that patient to the lesion into his globus pallidus or thalamus and the maintained relief of the symptoms may be tested over a period of days before any final destructive alcohol lesion is made. 2) The distention of the balloon for a certain period of time creates a small cavity where the alcohol may be injected without any undesired spreading along the needle tract.

STEP 10—A mixture of alcohol pantopaque is injected along the lumen of the needle after deflating the balloon within two days following surgery. We added pantopaque to our alcohol in order to be able to visualize the size and shape of the actual lesion and its progressive enlargements. Usually two to three injections are carried out over a period of eight to ten days the total amount injected usually being no more than 2 cc. and no less than 1 cc.

The following two cases are presented as examples of successful surgical intervention via chemopallidectomy in cases of dystonia musculorum deformans.

Case 1.

This 16 year old boy had been well until the age of seven when it was noted that the left foot became involuntarily inverted. It was soon apparent that the movements of the left foot were dystonic in nature. Within the next two years gross, deforming dystonic movements of all four extremities were noted and a conclusive diagnosis of dystonia musculorum deformans was made. Two years before admission to our service the patient became bedridden and the dystonic manifestations were so severe that both hips were chronically dislocated. The feet were markedly deformed and there was a very severe dystonic scoliosis Because of the marked deterioration of the patient's condition, with lack of useful voluntary movement of any extremity, accompanied by dystonic movements and fixed postures, and resulting total incapacitation, neurosurgical treatment was requested.

On October 25, 1955 a left chemopallidectomy was performed. This relieved the dystonic movements of the right extremities. Motor power

was preserved in these extremities so that after this procedure, the patient could once again feed himself and made considerable progress towards independence and self-care. On February 2, 1956, a right chemopallidectomy was performed, which relieved all dystonic manifestations in the left extremities. In addition to relief of the dystonic movements, there was marked alleviation of fixed postures in all four extremities as well as in the thoracolumbar spine. The patient was once again able to sit comfortably without support and within three months after the second operation, he was able to walk well with the use of two canes. These aids were necessary only because of the long-standing, chronic dislocation of the hips.

The patient has been seen for more than two years since the first chemopallidectomy and has had progressive improvement in all activities of daily living since that time. There are no longer any dystonic movements present. The patient attends a public school. Since the relief of the dystonic movements and postures has endured, orthopedic correction for the hip deformities is now contemplated. At the present time, the patient is completely independent in all activities of daily living.

Case 2.

This twelve year old girl had been well until the age of eight, at which time involuntary, twitching movements of the right hand were noted. Over the subsequent months, involuntary movements of a choreiform nature developed in all four extremities and were present at all times except during sleep. They gradually involved the nuchal musculature, and dystonic, writhing movements of the trunk developed as well. In 1953 a diagnosis of dystonia musculorum deformans was made, and this diagnosis was confirmed at various neurological clinics. By January, 1955, the involuntary movements and postures of all extremities were so severe that the patient was forced to discontinue attendance at school. Difficulty in feeding became marked so that her weight fell from a normal weight of 110 lbs. (50 kg.) to 80 lbs. (36 kg.). By the end of 1955, the involuntary movements were so violent that the patient had to be tied into a wheelchair or onto her bed to avoid her being thrown to the floor. Because of the desperate nature of symptoms at that time, operation was decided on.

Bilateral chemopallidectomy was performed on February 25, 1956. At that time, partial destruction of both the right and left globis pallidis was performed by injection of Etopalin, an 8% solution of a specially prepared cellulose in 95% ethyl alcohol.

This patient had a very uneventful convalescence. She was fully alert and well oriented the day after operation and was free of involuntary movements of all four extremities. The patient was out of bed on the fifth postoperative day and was able to sit in a chair without any assistance. She walked without assistance on the 7th postoperative day and was discharged on the 13th postoperative day.

Since the time of discharge from the hospital, twenty-two months ago, there has been continued improvement. The dystonic deformities of the

feet disappeared within the first three months. There are no involuntary movements in any extremity. The patient can walk, run, dance, and ride her bicycle. She once again attends public school. Relief of the dystonic movements and fixed postures has endured from the time of surgery until the time of this writing.

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CHAPTER XX

Surgery of Mental Illness

James W. Watts

Lobotomy has been performed in a relatively small number of children. The indications for operation are limited and the results are often disappointing. But there is the rare child who is so lacking in sense of personal preservation that he repeatedly injures himself and is so hostile, aggressive and destructive that facilities are not available to care for him, in whom lobotomy may offer the only solution. There may be others whose personality is undergoing steady deterioration in spite of psychiatric treatment in whom lobotomy may be considered.

Description of the Disease Entity

Most of the children treated by lobotomy exhibit a schizophrenic type of reaction. Schizophrenia in childhood is a malignant psychosis usually developing insidiously and progressing unchecked to complete decadence of the personality. It is frequently mistaken for mental deficiency but presents certain criteria that set it apart—an early period of normality or some precocious or peculiar skill. The outstanding feature in seclusiveness. Children who develop schizophrenia before puberty may show seclusiveness before the age of two. This gradually grows worse and efforts to bring the child into social relationship with others fail.

Another trait that may stamp the child as peculiar is the inability to react normally to praise or blame. There is no show of affection for others and a sullen hostility is shown toward those who look after them. Rages result from interruption of the child's preoccupation. Temper tantrums are brought out on slight provocation. These children do not play with toys, they use them as weapons or as objects to be torn apart or destroyed. If there are any pets in the household the child mistreats them.

Intellectually the children may appear precocious, but their precocity comes to be limited more and more to one particular category. Their personal habits become disorganized. While a child may have learned proper control at the usual age, there is a regression in toilet habits. Some children defecate in peculiar places or in their clothes, while others smear feces. Some collect trash, others eat it. Language is affected. An early sign of a

developing psychosis may be the peculiar use of words or peculiar pronunciation of them, which cannot be corrected. Some children are thought to be deaf and attempts are made at lip reading or speech correction, obviously without results. Language may become progressively less comprehensible with stereotyped phrases and eventually mutism. Hallucinations and delusional experiences are not so readily brought out in these children as they are in adults with schizophrenia.

Frequently the change in personality of the child is preceded by a febrile illness with drowsiness suggestive of central nervous system involvement, namely primary encephalitis or post-infectious encephalitis. In others, the alterations in behavior come on after a series of convulsive attacks. While encephalitis is difficult to prove, a post-encephalitic psychosis could explain the symptoms in some of our patients.

Technique

The standard Freeman-Watts prefrontal lobotomy was performed in the early cases. The pathways were sectioned in both frontal lobes in the plane of the coronal suture directly down to the sphenoidal ridge. This plane may pass a few millimeters anterior to the frontal horns of the ventricles or cut across them. When this operation failed to modify the behavior, a secondary operation, the radical lobotomy, was carried out cutting the pathways a predetermined distance posterior to the sphenoidal ridge. The distance ranges from 6 to 14 mm. These methods are described in detail in the second edition of *Psychosurgery* by Walter Freeman and myself.

Later in the series the radical lobotomy was employed as the operation of choice.

Numerous techniques may be employed but perhaps one of the better ones, selective destruction of the ventral medial quadrants of the frontal lobe by electrocoagulation, has been proposed by Grantham. He inserts insulated electrodes into the frontal lobe parallel to the sagittal sinus anterior to the frontal horn of the lateral ventricle which is roentgenographically outlined with a small amount of air. The position of the electrodes is checked by x-rays and if satisfactory they are withdrawn 1.2 cm. and cauterized. They are again cauterized after being withdrawn another centimeter.

Experience and Results

Prefrontal lobotomy has been undertaken in twelve individuals whose schizophrenic type of reaction developed before the age of ten. All of the patients were studied by Doctor Walter Freeman; three were operated upon by Doctor Jonathan M. Williams, and nine by the author. Ten children underwent operation between the ages of four and a half and fourteen years, and two at the age of fifteen. Most of these twelve patients had received the

benefit of prolonged and careful observation in recognized institutions, and came from homes where facilities were available for satisfactory after care. Parents were still sufficiently interested in them to make very considerable sacrifices in their behalf during the difficult period of training that followed operation.

Lobotomy may seem radical until one has seen these children and talked to their parents. A few case histories will illustrate some of the problems.

Case 1

In 1943, a physician brought his four and a half year old son to my office. The boy's face was a mass of bruises as a result of banging his head against furniture in response to the most trivial frustration. While I was taking the history, he escaped from his parents' clutches, banged his head against the radiator and, before we could restrain him, had knocked his head against the window. He let out a yell each time he struck himself. When restrained, he screamed as if he were being murdered. This was typical of his behavior. He was destructive, assaultive and absolutely incorrigible. The boy never talked, had not said "Daddy" or "Mama." Unfortunately the possibility in this case will remain unknown since after his return home, and when things were going well, he contracted meningitis and died three weeks after operation. Necropsy showed clean healing wounds.

Case 2

This little girl was first seen in August of 1944 at the age of six. She walked and talked at twelve months and had control of her bladder at two years. She had an ordinary attack of measles and chicken pox when she was three and a half years of age with no complications. The onset of her symptoms was gradual, characterized by destructive behavior, screaming spells and insomnia. She never made friends with children, spent long periods in the bathroom taking two to three baths a day, washing her hair repeatedly. She stopped talking when she was three. The child was examined at the Duke University Hospital at the age of four when an electroencephalogram showed generalized dysrhythmia and a diagnosis of post-encephalitic syndrome was made. She was seen again at Duke in 1944 where a Stanford-Binet test showed her I. Q. to be 34.

When we saw her in 1944 she tore her clothes, smashed her toys and used the toys as weapons. She was admitted to the George Washington University Hospital where a pneumoencephalogram was normal. A prefrontal lobotomy was carried out in August of 1944 and she improved for about two months, slept better, was less destructive but relapsed within a few months. A second lobotomy was performed in April of 1945, 15 mm. behind the previous one. Recovery was slower following the second operation. During the next year she was less destructive, began putting a few words together and made a few sentences. She remained at home until 1950 when she was admitted to the Caswell Training School.

In a study carried out with Newman and Hohman at Duke University, Doctor Hal C. Holland interviewed the girl at the training school. He was unable to obtain any verbal response in answer to questions but she seemed happy in her environment. She was able to feed and dress herself and was tidy in her toilet habits. She was examined at the school in November of 1956 by Doctor Walter Freeman. He reported that she seldom utters a word. She does not participate actively in any of the work of the children. She has occasional convulsive attacks.

Case 3

In 1949 Doctor Williams presented a nine year old boy at the Post-Graduate Course in Psychosurgery held by the George Washington University School of Medicine. His college educated parents noted precociousness in the child until the age of three and a half when he was ill with a high fever and drowsiness for about a week. Following this seige he showed progressively intolerable behavior, being destructive, hostile and aggressive. He even dived through closed windows. Toilet training was lost. For a period, to prevent harm to himself and others, he had to be confined in a cage in the basement where he was fed through a small trap door in the wall by his sorrowing parents.

On July 31, 1957, I talked with the patient's mother who informed me he is living at home. He never loses his temper and has had no temper tantrums. She describes his disposition as "sweet" and says that he is neither destructive nor aggressive. He has good control of his bladder. He dresses himself, has learned to tie shoes, and is quite neat. He rarely initiates a conversation but answers questions about things that are familiar to him. He knows his address, knows the members of his family and where they live, can write his name and address. He likes music, listens to radio but does not care for television. His mother thinks that the latter is because his vision is poor.

The patient is being followed at the District of Columbia General Hospital and is taking small doses of Thorazine.

A seven to seventeen year follow-up study in our series of twelve children reveals that two are dead, six are in special schools or mental hospitals, and four are living at home. None has been rehabilitated. On the other hand, there has been some improvement in behavior as illustrated by Cases 2 and 3.

Doctor Hal C. Holland, who is now Senior Resident in Neurosurgery in the George Washington Univeristy Hospitals Program, allowed me to review a study which he made with Newman and Hohman while at Duke. In the Duke University series of lobotomized children, the most striking finding was the reduction of destructive tendencies by lobotomy. Ten of eleven patients were destructive before operation and none have destructive tendencies at the present time.

In contrast to adults who receive the maximum benefit from psychosurgery within six months of operation, they found that children may go one to three years before they reach a turning point for the better.

When Doctor Freeman and I wrote the Second Edition of Psychosurgery in 1950, we included the children in the chapter on Schizophrenic Reaction Type. Some of these children were seclusive, most exhibited some peculiar traits and had temper tantrums on the slightest provocation. All but one were behavior problems. Two had never learned to talk but appeared quite alert and had good coordination and normal reflexes. Most of the lobotomies were done in children between 1939 and 1948. We believe that the symptoms were primarily due to emotional tension and the proper classification was schizophrenia. After reviewing the records in preparation for this chapter with the advantage of a follow up ranging from seven to seventeen years, it seems probable that many of these children had organic brain disease. The parents date the behavior change to a febrile illness or a series of convulsions in an appreciable percentage. It also seems probable that the failure of two of the children to develop speech was due to organic disease rather than to emotional conflicts.

The indications for lobotomy in children are limited and results are disappointing. Operation is most effective in reducing hostility, aggressiveness and destructive tendencies.

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CHAPTER XXI

Surgery of Epilepsy

MAITLAND BALDWIN

EPILEPSY is a clinical entity. It is characterized by recurrent, paroxysmal alterations of consciousness, unusual movements, and bizarre sensations. These phenomena usually constitute the outward expressions of a cerebral seizure.

A cerebral seizure is actually an abnormal neuronal discharge, which usually spreads over normal pathways. It originates within groups of nerve cells which have been altered so that their intrinsic chemistry is abnormal. These cells have an abnormal potassium, glutamine, and asparagine metabolism. Usually the surrounding glial or non-neuronal cells are also changed and these changes are discernible in histological preparations as areas of gliosis or sclerosis.

This aggregate of abnormal cells constitutes an epileptogenic lesion. Such a lesion can occur as the result of neoplasm, trauma, abscess, or more generalized infection and various anoxic changes. In children it may also occur along the frontiers of an area deficient in vascular supply because of congenital or acquired disease. It always occurs along the frontiers of the precipitating or etiological lesion, never within the precipitating lesion itself.

Epileptogenic lesions cause recurrent paroxysms or cerebral seizures. If the causal factor which precipitated this neuronal lesion is neoplastic, the paroxysms will change as the new growth progresses. However, if anoxic insults precipitate epileptogenic lesions, these cause habitual or stereotyped seizure patterns which do not change as the clinical entity develops. Of course any cerebral neuronal system can discharge in an epileptiform manner, but this is not necessarily indicative of an epileptogenic lesion. Rather, it indicates a fundamental property of otherwise healthy human neurones. Thus, under certain circumstances, almost any human cerebral system (of neurones) can discharge in an epileptiform pattern. For example, transient hypoglycemia, uremia, chemical intoxication (lead, strychnine), pyrexia, or hydration may provide these unusual circumstances Likewise, analeptic drugs such as Metrazol, or electrical currents as used in psychiatry, may produce epileptiform discharges and

cerebral seizures. These seizures seldom recur, while those caused by an epileptogenic lesion usually recur and become habitual.

Most cases of habitual cerebral seizures begin in childhood. Certainly an epileptogenic lesion may develop at any age, but most frequently these lesions develop in the early years of life. In the majority of the 1,500,000 epileptics (in the United States today) the onset of attacks occurred before the age of twenty years. Actually, many individuals experience their first seizures in infancy or childhood. However, very few of those afflicted with habitual cerebral seizures develop them because of hereditary factors. Approximately 0.1% of all cerebral seizures can be related to hereditary factors.

It seems clear that the problems of epilepsy must be encountered by those who care for sick children. These problems are always manifold. There are always medical, psychological, and social aspects. Occasionally the ultimate problem can only be solved by surgery. Certainly no solution is adequate unless it includes practical considerations of medical, psychological, and social characteristics. Likewise, surgical treatment of epilepsy is never successful unless it is preceded by practical consideration and adequate analysis of the various aspects.

The Medical Problem

The medical considerations can be subdivided into diagnosis, classification, and therapy.

Diagnosis

Diagnosis must begin with a careful history. Each history should contain an accurate description of the habitual seizure, and each seizure description must include evidence derived from the patient and relatives. The patient may have a warning of the approaching seizure and this is most important, for the nature of the warning may provide an invaluable clue to the origin of the attack within the brain as shown in Figure 1. Paradoxically, the absence of a warning may also be important in localization. In all cases, the first or initial phenomena are the most valuable for observation and diagnosis These first phenomena often indicate the origin and immediate direction of the abnormal neuronal discharge. Conversely, the final phenomena in each habitual seizure are usually the result of diffuse spread and therefore have little value in determination of origin and localization within the brain.

An accurate history of the onset, sequence, and end of each seizure usually provides the first available evidence for localization of the epileptogenic lesion. This is the first step in diagnosis. It must be followed, whenever possible, by direct observations of the spontaneous habitual seizure. Such observations should be recorded accurately because the events of

the clinical seizure are indicative of the origin and spread of the neuronal discharge. Thus, as the abnormal electrical activity originates, it activates certain functional representations and it continues to activate others as it spreads over the adjacent neuronal circuits. Finally, it may activate many circuits and cause a generalized seizure. Its action is always mirrored by the clinical phenomena, whether these are bizarre sensations, alterations in consciousness, or unusual movements. Actually, each clinical seizure is a highly organized series of events, and each element or event is indicative

Initial Phenomena	*Number of Cases*	*Localization*
INITIAL UNCONSCIOUSNESS	30	Chiefly frontal (20% temporal)
MOTOR PHENOMENA	29	
Jacksonian motor	20	Rolandic area
Vocalization	2	Intermediate precentral
Simple adversive	7	Chiefly intermediate precentral
SENSORY PHENOMENA	81	
Sensory somatic	55	
Sensory visual	11	Chiefly Rolandic area
Sensory auditory	3	Occipital
Sensory vestibular	9	Temporal
Sensory olfactory	1	Temporal and parietal
Sensory gustatory	2	Uncus
AUTONOMIC MOTOR AND SENSORY PHENOMENA	27	Sylvian
Epigastric and abdominal sensation	8	Chiefly Sylvian region and intermediate precentral
Epigastric rising sensation	8	Intermediate precentral
Palpitation (epigastric and precordial)	6	Intermediate precentral
Thoracic sensation	3	Sylvian region
Miscellaneous autonomic phenomena	2	?
PSYCHICAL PHENOMENA	29	
Hallucinations	5	Temporal
Illusions	13	Temporal and frontal
Fright	3	?
Forced thinking	2	Frontal
Aphasia	6	Frontal & temporoparietal
MISCELLANEOUS	26	
Body sensations	9	Frontal, parietal, temporal
Cephalic sensations	16	Frontal, parietal, temporal
Yawning	1	Frontal?
TOTAL	222	

FIGURE 1. Initial phenomena and cerebral localization (Penfield and Kristiansen, 1951).

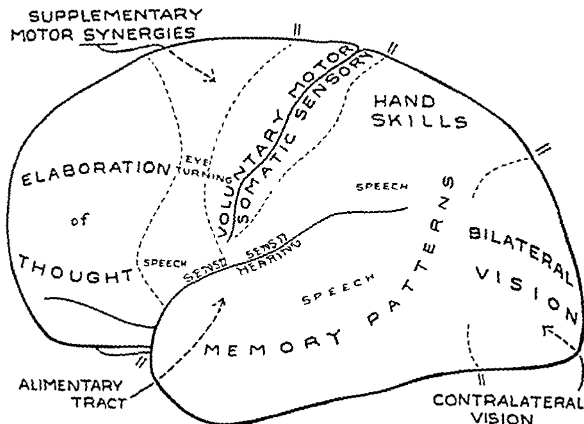


FIGURE 2. Summary of major functional representations of the human cerebral cortex (Penfield and Rasmussen, 1950).

of the part or parts of the brain most affected by the abnormal discharge.

Since the elements of clinical seizures can be recognized as distortions of normal functions, it follows that a reasonable knowledge of functional anatomy provides the basis for seizure analysis. Some of the pertinent functional anatomy is summarized in Figure 2.

Localization

The various functional representations in these figures may be activated by a seizure discharge so as to produce phenomena which are recognizable in terms of their origin. An epileptogenic lesion close by or in the precentral cortex might discharge so as to activate motor phenomena. These will occur in sequence comparable to the spread of the discharge across the motor cortex. Similarly, a discharge could activate the sensory cortex so as to produce a sequence of sensations which the patient could describe and remember. Epileptic discharge beginning in either motor or sensory cortex is usually followed by sensations or movements. In other words, motor and sensory cortices are seldom activated alone but rather in concert. The aftermath of such discharge can be analyzed by clinical examination. There is usually a postseizure weakness or hypesthesia related to the areas of the cortex most affected by the discharge. This postictal weakness is most frequent

after seizures originating in the motor cortex and most frequent in children. It is a valuable localizing sign, which may be accompanied by an extensor plantar reflex and absent abdominals. A well-localized cortical lesion causes focal cerebral seizures. Such lesions usually follow penetrating wounds of the brain and occur along the frontiers of the resultant meningo-cortical cicatrix.

Focal epileptogenic lesions elsewhere in the cortex produce clinical seizures with different characteristics. These may be understood after careful study of the previous figures. However, certain focal cerebral seizures have particular clinical characteristics which require special emphasis. Thus, an epileptogenic lesion close by a functional representation for speech does not activate speech function; it arrests speech. Such a speech arrest or dysphasia may be the first event in the seizure pattern or it may occur somewhat later in the ictal sequence. When it occurs as an ictal event, it usually continues in the postictal period. Likewise, a focal discharge in the frontal cortex causes an immediate alteration of consciousness, and initial alteration of consciousness is indicative of frontal origin or origin deep within the brain stem.

Finally, an epileptogenic lesion in one or the other temporal region usually produces bizarre psychological changes and unusual mental experiences, as well as certain motor and autonomic phenomena. These are pallor, pupillary dilatation, movements of face and jaws, inspiratory arrest, and salivation. Certain warnings or aurae are considered characteristic of origins in the temporal lobes, and these are listed in Figure 1. Epileptic discharges in the temporal lobe also seem directly related to unusual behavior consisting of alteration of consciousness, searching movements, and apparently purposeless activity. Temporal lobe seizures are characterized by bizarre behavior. However, the bizarre ictal behavior is no more striking than the unusual behavior which characterizes the periods between clinical seizures. Thus a child with an epileptogenic lesion in one or the other temporal lobe may be aggressive and destructive, or he may develop otherwise unexplained outbursts of temper. Frequently this antisocial behavior becomes a serious problem for parents and teachers. Then the child is brought to the physician as a problem in behavior, rather than a clinical problem in the control of seizures. An adequate historical analysis, supplemented by observation of the seizures, provides the essential diagnosis in these so-called "problem children."

Psychological Testing

After accurate recording of the seizures and adequate clinical examination the child should receive the benefit of psychological testing and psychiatric assessment. Some children with cerebral seizures are retarded;

others have subtle deficits in speech which make learning difficult. The degree of mental retardation and the character of speech deficits must be estimated in order to provide a realistic background for future treatment. Certainly those children with congenital lesions of the brain are most likely to have some mental retardation. Similarly, any epileptogenic lesion close by an area of speech representation will cause chronic deficits in communication. These must be noted during diagnostic examination and assessed when treatment is considered. Furthermore, the behavior of the child must be assessed in terms of its individual significance and its significance within the family. The parents and the siblings should be interviewed so as to determine their emotional relationship to the afflicted child. Frequently, the parents require more supportive therapy than the patient. Furthermore, treatment of the epileptic child cannot be adequate or efficient unless the family situation is understood and evaluated. Actually an epileptic child often constitutes a "social abscess" within the family structure. This tragic relationship can be avoided or beneficially altered by the responsible physician. He must learn to recognize its characteristics in the first interviews and examinations so that the subsequent treatment will provide for future alteration.

The history, seizure observation, clinical examination, and psychological evaluations constitute the primary steps in diagnosis of cerebral seizures. The seizure history and observations may indicate that the attacks begin without warning and continue as generalized phenomena. In such cases the clinical examination must be reinforced by laboratory assessment of fasting blood sugar, blood calcium, and kidney function. Likewise, the historical data must be searched for rare etiological factors such as heavy metal poisoning or unusual infections. Actually, many generalized seizures owe their origin to a lesion of unknown etiology deep within the brain. However, the rare etiological factors sometimes play a significant part. The primary clinical and neurological examination must emphasize lateralizing signs. A relative smallness, unilateral weakness, or abnormal reflex may lateralize the epileptogenic lesion within the brain. Certainly lateralizing signs are most frequently observed in the postictal period. Therefore, no clinical examination is complete unless it includes observations after an attack. During this period aphasic errors may occur. These are signs of a left-sided lesion in a right-handed child and vice versa.

After the primary steps in diagnosis have been completed, the secondary or corollary steps must be taken. These are electrographic and radiographic examinations. These tests should be guided by information derived in the primary examinations.

The electrographic recordings are invaluable in most cases, but their value cannot be fully realized unless based on a shrewd clinical diagnosis.

The electroencephalographer should be told "where to look." He may prove the clinician wrong in some cases, but in most cases his evidence must be substantiated by clinical observation.

The electroencephalographer looks for certain characteristics in his recordings. These are illustrated in Figure 3. He may obtain this type of evidence in one scalp recording or it may not be found in many. Frequently he will use various electrode placements or arrangements. Among these are pharyngeal electrodes designed to provide special recordings from the temporal fossae. In children any recording requires sympathetic explanation as well as deft technique. Some recordings will be assisted by injection of barbiturates. These produce "sleep tracings" during which temporal lobe lesions may be discovered. Occasionally the recording may be "activated" by an injection of Metrazol. This technique is particularly indicated when several normal or borderline tracings have been obtained. The injection of Metrazol may be useful for its effect on the tracing or because it will produce a clinical seizure. Such a seizure is often quite comparable to the spontaneous attacks.

The radiographic examination should include plain films of the skull in anteroposterior and lateral projections. These should be taken so as to provide for stereoscopic analysis. Frequently the x-rays demonstrate a relative smallness of one-half of the calvarium. This may be associated with elevation of the ipsilateral middle fossa and thickening of the parietal eminence. These signs indicate a relative smallness of one hemisphere, and the smaller hemisphere may contain the epileptogenic lesion. Certainly the plain films must be carefully searched for evidence of abnormal intracranial pressure or intracerebral calcification. Widening or separation of the sutures usually denotes increased intracranial pressure in childhood. It occurs long before the dorsum sella can be decalcified. Likewise, intracerebral calcification may indicate a neoplasm. When the intracranial pressure is normal, a pneumographic study is often indicated. This may demonstrate a relative dilatation of the ventricular system or focal dilatation of one of its parts. Either finding may indicate the side or location of a significant lesion. Obviously, cases with increase of intracranial pressure should be studied by ventriculography. Arteriography may be indicated if a vascular malformation is suspected.

Classification

The electrographic and radiographic evidence must be correlated with the clinical data. Such correlation provides the basis for a reasonable classification of seizures. This classification begins with a consideration of the pertinent functional anatomy and is elaborated by combination with relevant electrographic data. Thus, seizures characterized by certain initial

events followed by a recognizable sequence can be interpreted in terms of the functional anatomy, e.g., an attack beginning with a twitching of the left thumb and continuing with clonic movements of the left upper extremity. This attack could be interpreted as beginning in the right motor cortex. Its continuation involved the motor cortex on the right side. Therefore, it is a "focal cerebral seizure, right somatomotor." If the seizure cannot be localized in terms of functional anatomy, it may be called simply a "cerebral seizure, unlocalized." By custom, certain electrographic characteristics provide a subclassification. This is the spike-and-wave complex (Figure 3). When this occurs as the outstanding electrographic characteristic, the seizure is called "centrencephalic" (Figure 3a). This word is used

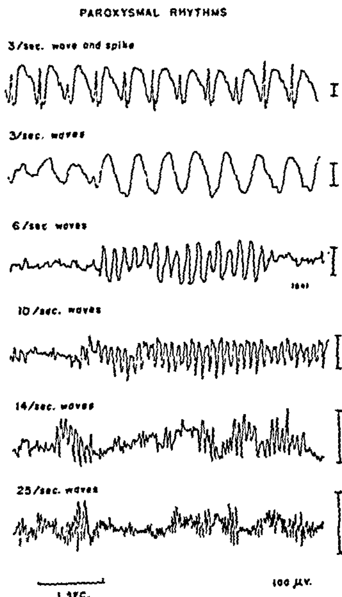


FIGURE 3. Electrographic records characteristic of certain types of epilepsy: (a) "centrencephalic" discharges (Penfield and Jasper, 1954).

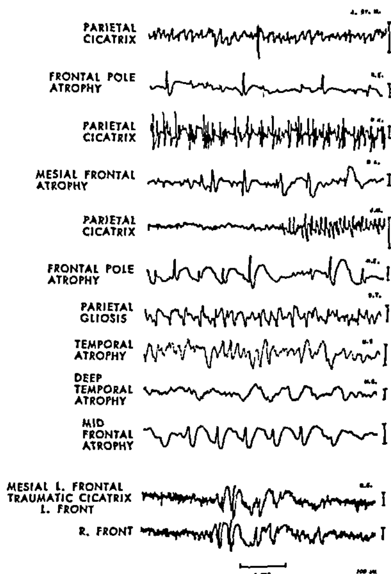


FIGURE 3. Electrographic records characteristic of certain types of epilepsy: (b) "focal" discharges (Penfield and Jasper, 1954).

to denote its supposed origin deep within the brain stem. It is equivalent to the term "petit mal."

During classification of the actual seizure, the physician must also consider its immediate effects. Focal cerebral seizures in children may be followed by serious motor, sensory, or language deficits. These postictal changes may be irreversible if the seizures continue uncontrolled for long periods Certainly, repeated severe attacks of any type may be followed by serious changes. Indeed, a young patient with centrencephalic seizures may die during or after a period of uncontrolled attacks. Yet the most frequent and most serious postictal changes occur after focal cerebral seizures.

Medical Treatment

All changes in medication should be gradual. The dosage should be gradually increased to the limits of toxicity, but initial large doses should be avoided. Likewise, a change in type of medication should be a process of slow withdrawal and gradual substitution. Some combinations are effective, i.e., Phenobarbital and Dilantin, and two or more preparations may be used together. However, each drug in the combination should be first prescribed in small doses. There are certain times when a temporary increase in dosage is necessary. During periods of fever, menstruation, hydration, or constipation, the dosage should be increased. Conversely, it may be necessary to withdraw medication for purposes of complete electrographic examination. This withdrawal should be accomplished in hospital because of the danger of status epilepticus. In this event the child should be promptly anesthetized. Anesthesia should be continued at a surgical level for at least one hour. Certainly the administration of a general anesthetic to children may be regarded as a serious or even dangerous undertaking. However, the serious and dangerous consequences of status epilepticus far outweigh the minimal risks of anesthesia administered by competent hands.

There are three generalizations which may be useful in any consideration of anticonvulsant medication. Change is inevitable, but it must be gradual. Withdrawal may be necessary, but it is dangerous. Dosages are increased so as to approach the limits of toxicity, but the signs of toxicity must be anticipated. Figure 4 provides a list of useful anticonvulsant preparations, with suggestions on initial dosage. Figure 5 lists the drugs, with the comparable toxic reactions.

Certain drugs, such as Phenobarbital, Dilantin, and Mebaral, may be considered as having various applications. Others, such as Tridione and Paradione, are most applicable to centrencephalic disorders. All anticonvulsant preparations should be carefully considered in most cases.

The application of these drugs is only a part of medical treatment. The general principles of pediatric care cannot be overlooked, and one must always consider the psychological and social factors as well. Diet is important. Some have recommended ketogenic combinations of food values. In all diets excessive hydration must be avoided. Also, dietary regimes must be controlled so as to prevent constipation. Both hydration and constipation may be associated with severe attacks. Finally, the personality, intellect and behavior can be altered by anticonvulsant medication when this approaches the limits of toxicity. Thus many anticonvulsant preparations seem to depress intellectual activity, and an otherwise intelligent child may appear retarded when he receives high doses of medication. Likewise,

FIGURE 4. Average starting and maximum dosages* of anticonvulsant drugs** used in the treatment of Epilepsy (Livingston, 1954).

Drug	Age (Years)	Starting Dosage		Maximum Dosage	
		Metric	*Apothecary*	*Metric*	*Apothecary*
Bromides	under 6	320 mg 2x daily	5 grains 2x daily	640 mg. 3x daily	10 grains 3x daily
	over 6	320 mg 3x daily	5 grains 3x daily	1.0 Gm. 3x daily	15 grains 3x daily
Phenobarbital	under 3	16 mg. 3x daily	¼ grain 3x daily	32 mg. 3x daily	½ grain 3x daily
	3 to 6	32 mg. 2x daily	½ grain 2x daily	65 mg. 3x daily	1 grain 3x daily
	over 6	32 mg. 3x daily	½ grain 3x daily	100 mg. 3x daily	1½ grains 3x daily
Mebaral	under 3	32 mg. 3x daily	½ grain 3x daily	65 mg. 3x daily	1 grain 3x daily
	3 to 6	65 mg 2x daily	1 grain 2x daily	130 mg. 3x daily	2 grains 3x daily
	over 6	65 mg 3x daily	1 grain 3x daily	200 mg. 3x daily	3 grains 3x daily
Dilantin	under 6	32 mg. 3x daily	½ grain 3x daily	100 mg. 3x daily	1½ grains 3x daily
	over 6	100 mg. 2x daily	1½ grains 2x daily	200 mg. 3x daily	3 grains 3x daily
Mesantoin	under 6	50 mg. 3x daily	¾ grain 3x daily	200 mg. 3x daily	3 grains 3x daily
	over 6	100 mg 3x daily	1½ grains 3x daily	400 mg. 3x daily	6 grains 3x daily
Benzedrine Sulfate	under 6	2.5 mg. 2x daily		5 mg. 3x daily	
	over 6	5 mg. 2x daily		15 mg. 3x daily	
Dexedrine Sulfate	under 6	2.5 mg. daily		2.5 mg. 3x daily	
	over 6	2.5 mg. 2x daily		7.5 mg. 3x daily	
Tridione	under 6	150 mg. 2x daily	2¼ grains 2x daily	300 mg. 3x daily	4½ grains 3x daily
	over 6	300 mg 2x daily	4½ grains 2x daily	600 mg. 3x daily	9 grains 3x daily
Paradione	under 6	150 mg. 2x daily	2¼ grains 2x daily	300 mg. 3x daily	4½ grains 3x daily
	over 6	300 mg. 2x daily	4½ grains 2x daily	600 mg. 3x daily	9 grains 3x daily
Phenurone	under 6	250 mg. 3x daily	3¾ grains 3x daily	1.0 Gm. 3x daily	15 grains 3x daily
	over 6	500 mg. 3x daily	7½ grains 3x daily	2.0 Gm 3x daily	30 grains 3x daily
Gemonil	under 6	50 mg 3x daily	¾ grain 3x daily	100 mg 3x daily	1½ grains 3x daily
	over 6	100 mg 3x daily	1½ grains 3x daily	200 mg. 3x daily	3 grains 3x daily

* The conversion of the metric system of weights to the apothecary system is approximate.
** This table includes only those drugs which were available for general usage at the time of this writing (1954).

FIGURE 5. Common untoward reactions of anticonvulsant drugs (Livingston, 1954). This table lists the common and significant untoward disturbances which have been observed during the course of administration of anticonvulsant drugs. Some of these reactions are indicative of an overdosage of the drug; others represent sensitivity reactions and some are toxic manifestations.

Name of Drug	*System Involved*					
	Nervous	*Cutaneous*	*Gastro-intestinal*	*Hemopoietic*	*Genito-urinary*	*Cardio-vascular*
Bromides	Drowsiness	Acneiform eruptions (Only in adolescent children)	None*	None*	None*	None*
Phenobarbital and Mebaral	Drowsiness Increased excitability	Rare**	Rare**	None*	None*	None*
Dilantin	Ataxia Diplopia Nystagmus	Rash (scarlatiniform or measles-like) Hyperplastic gums Hirsutism	Nausea Vomiting Constipation	Rare**	Rare**	Rare**
Mesantoin	Drowsiness	Rash (scarlatiniform or measles-like) Purpuric rash Exfoliative dermatitis	Rare**	Agranulocytosis Pancytopenia	None*	None*
Benzedrine and Dexedrine Sulfate	Increased irritability or restlessness Insomnia Tremor	None*	Anorexia Loss of weight	None*	None*	None*
Paradione	Photophobia	Rash (various types)	Hiccough	Agranulocytosis Pancytopenia Thrombocytopenia	Rare**	None*
Tridione	Photophobia	Rash (various types)	Hiccough	Agranulocytosis Pancytopenia	Rare**	None*
Phenurone	Alterations in behavior and personality Headache Insomnia	Rash (scarlatiniform or measles-like)	Hepatitis Anorexia Nausea Vomiting Abdominal pain	Leukopenia Pancytopenia	Transient albuminuria	Rare**
Gemonil	Drowsiness	Morbilliform rash	None*	None*	None*	None*

mentally retarded children may suffer further depression of intellectual activity because of medication. Obviously, these children with mental defects require special training and cannot compete in usual school environments. Moreover, the parents of epileptic children require attention, as theirs is a tragic burden often increased by their own emotional reactions.

These unhappy children are best treated by the combined efforts of physicians and social workers. An adequate therapeutic combination usually requires coordination of the pediatrician, psychiatrist, and social worker. Indeed, some cases may also require the assistance of a neurosurgeon.

The Surgical Treatment

Surgery offers definitive and effective treatment in certain cases. The *focal cerebral seizures which do not respond to medical therapy should* be assessed for radical intervention. In general, each case must be given a

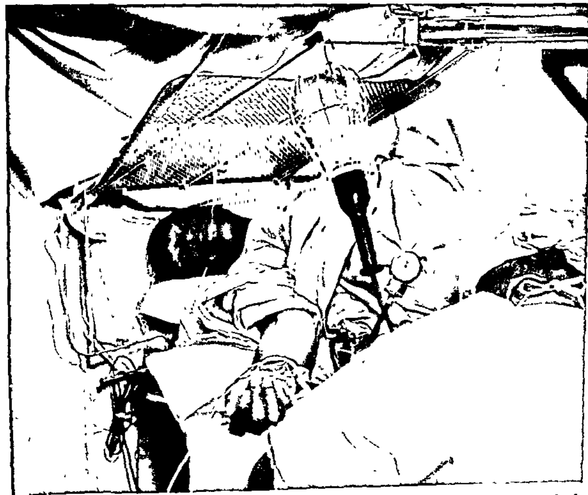


FIGURE 6. The patient during electrical exploration of the cortex. Note draping which shields surgical field and exposes patient for observation; also small black microphone close to patient's mouth so surgeon and others may clearly hear patient's response to stimulation.



FIGURE 7. Cortical electrodes placed on the surface of exposed cortex and attached to EEG machine in an adjacent room. The electrodes are moved about entire exposed area for epileptogenic foci. (*Courtesy of* Dr. Ira Jackson)

trial period of one year on medication. No case should be considered for surgery unless this period has been used for an intelligent application of various drugs. After this period the case must be analyzed by clinical, electrographic, and x-ray techniques so as to determine the localization of the epileptogenic focus. If an essential epileptogenic process can be localized by these techniques, the case may require operative treatment. Obviously, those cases with neoplastic disease require immediate surgical intervention. Moreover, if neoplasm is suspected (as the etiological factor), one cannot wait for the trial of anticonvulsant medication. Otherwise, there are three criteria for surgical treatment of epilepsy in children. The first is the failure of an adequate medical regime. The second is the adequate localization of the epileptogenic process. The third is the age of the child. Young children or infants should not be subjected to surgery for seizures. Localization is difficult in the early age groups and the surgical risk is great Arbitrarily, one might select children older than twelve years. In all cases the localization problem requires painstaking, repeated clinical and electrographic examination for adequate solution. This implies a period of at least two weeks' observation in hospital, during which the clinical

and electrographic examinations can be repeated. Habitual and activated seizures must be observed in this period of study. It should also include psychological testing, and it must end with a correlation of the data. The data derived from the clinical and electrographic examinations must indicate the same origin for the habitual seizures. If these data cannot be correlated to this end, surgical intervention is useless.

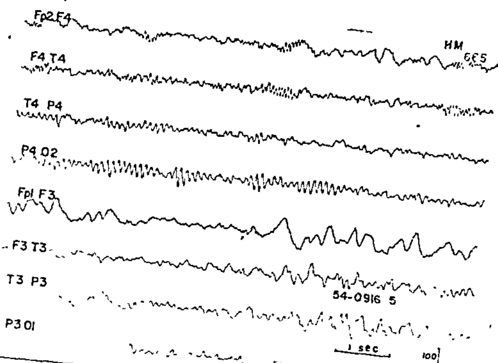
Once localization of the epileptogenic lesion has been accomplished, surgery can be considered. If the significant lesion is localized in an area related to speech or normal finger movement, it should not be excised. The functional loss after such excision far outweighs the therapeutic gain. However, focal lesions elsewhere can be excised with relatively little functional loss and reasonable expectation of therapeutic gain.

The therapeutic effect of operative treatment depends on the accuracy of localization and the precision of excision. At the operating table the lesion can be localized by electrical stimulation and electrographic recording. The surgeon must try to reproduce the aura or pattern of the habitual seizure by gentle electrical stimulation of the exposed cortex. It is always desirable and sometimes necessary to reproduce the initial phenomena of the attack. However, the surgeon must stimulate with care so as to prevent activation of a generalized seizure. If this occurs, the patient is endangered and further recording from the exposed brain is difficult or useless because of the postictal effects.

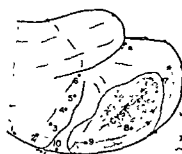
Electrographic recording should be made from all areas exposed. The localization of the lesion can be predicted by recording of epileptiform activity. Usually this is maximal over the lesion. Excision may be planned so as to include the areas from which the seizure phenomena have been reproduced or recorded. Actually, the results of excision will be most promising if the area removed also includes evidence of pathological change. Such evidence may take the form of microgyri, yellow color, change in

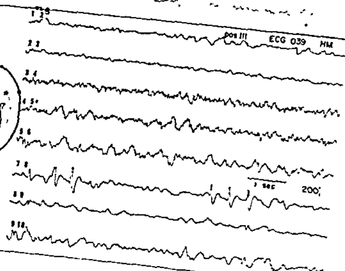
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FIGURE 8 (a). Preoperative tracing of 13-year-old boy whose seizures began with tingling in the right cheek which spread to the right arm. This sensation was followed by alteration of consciousness and posturing of right extremities. He had relative smallness of the right side and a right hemiparesis. Note asymmetry in record, as well as irregular slow waves on the right (Fpl F3) (*Epilepsia*, Series III, 1955, Vol. 4). (b) Electrocorticogram of case described in 8a. The position of recording electrodes and the extent of the cortical pathology are shown in the left hemisphere. Epileptiform discharges appear at borders of the cyst (*Epilepsia*, Series III, 1955, Vol. 4). (c) Electrocorticogram (after cystic lesion and part of the surrounding tissue have been excised (see Figure 8b). Electrode placements are shown in drawing above. A—Actively firing spikes remain in a relatively discrete area of excision's posterior frontier (Electrode 9). B—After further cortical excision (outlined by dotted line), the epileptiform activity disappears (*Epilepsia*, Series III, 1955, Vol. 4).

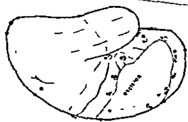


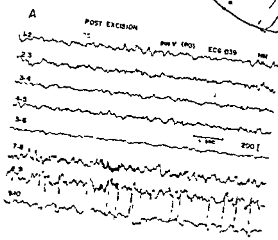
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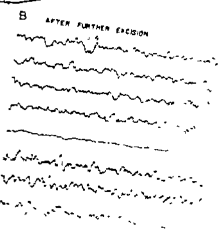


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POST EXCISION
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B
AFTER FURTHER EXCISION

consistency, or thickening of the overlying pia arachnoid. If the electrographic and objective evidence is equivocal, no excision should be made. On the other hand, excision will be advantageous if this evidence points to one area. After excision, further recordings may show a relatively normal record or, conversely, the persistence of epileptogenic abnormalities. If the abnormalities persist, further excision must be considered. Removals in the temporal lobe should include the lateral extent from anterior pole to the vein of Labbe. Such a removal should also extend mesially to the pia so as to include uncus and amygdaloid nucleus. Further recordings should then be made in order to determine the need for excision of the pes hippocampus and anterior hippocampal gyrus. Excision of the posterior-mesial structures is best avoided if bilateral temporal removals are contemplated.

Electrical localization is most important in surgery for epilepsy. Its success is dependent upon skillful application of the various electrodes and experienced interpretation of the results of stimulation and recording. These methods are most effective when the patient is under local anesthesia. Certainly local infiltration is a problem in pediatric surgery. Thus, the child may not be able to cooperate because of apprehension. In some cases this can be allayed by careful psychological preparation. Thereafter the brain exposure can be accomplished with nitrous oxide as a supplement to the local infiltration. However, when nitrous oxide has been used, recording must be delayed until all the gas has left the patient's system, as its presence alters the electrographic tracing. Some children cannot cooperate during the necessary period of wakefulness after the brain exposure. In these cases the operation must be done under general anesthesia with intratracheal intubation. In some such patients the general anesthesia may be supplemented with succinyl choline. Then the anesthetic may be discontinued while respiration is maintained artificially. This provides the necessary level of brain function and immobilization of the patient. The child is not uncomfortable if respiration is skillfully maintained, and anesthesia may be reinstituted after localization has been completed.

Excision must be precise in all cases. A careful subpial dissection with suction provides the most precise excision technique. This method permits adequate removal of the affected area. It also leaves intact pia on the margins of the excision. Thus the blood supply of the frontiers remains essentially intact after removal.

It seems clear that craniotomy for epilepsy requires specialized electrical and surgical techniques. These techniques are time-consuming and sometimes tedious. Therefore they should be applied in an operating room environment which can support long procedures.

The surgeon who would treat epileptic children must utilize facilities which provide opportunity for thorough postoperative study as well as

prolonged operative treatment. Surgical treatment is quite effective when the conditions of selection and operation are controlled. Under such conditions, carefully selected cases can be successfully treated. Certainly no case should be selected for radical treatment unless medical therapy has failed to control the attacks. Moreover, medical treatment must continue for at least a year after surgical therapy.

In any case, the medical or surgical therapy is only a partial solution. Psychological and social factors must form a reasonable portion of the total therapeutic regime. All therapy must be based on direct and electrographic observation of the habitual seizure patterns as well as psychological assessment of the afflicted child. These children are afflicted with a paroxysmal disorder, but the periods between paroxysms are fraught with psychological hazards.

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* Rather than a long bibliography of published articles in journals, these book references will inform the reader on this subject.

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